Part 2 (pages 112-236)

"We Can't Kill Your Mother!"
and Other Stories of Intensive Care

TABLE OF CONTENTS

14. Cocaine Wins

Lester Brown was a large man, at least 6 feet 2 inches and 240 pounds. Even asleep he looked menacingly big, someone you didn't want to wake up before he was ready. The day he was brought to the Emergency Department you could have shook and pinched and tickled Mr. Brown and he wouldn't bother you. He had a ruptured blood vessel at the base of his brain.

It started at home. A few minutes after snorting some cocaine his posterior communicating cerebral artery began to pulsate, causing him to complain of "the worst headache of my life." Seconds later he fell to the floor, unconscious. En route to the emergency department the artery burst and he suffered a full blown *subarachnoid hemorrhage*. The loss of an ounce of blood into the surrounding brain tissue relieved the arterial pressure and the hemorrhage ceased, but by then it was too late. Irritating blood washed over the normally smooth brain surfaces where it didn't belong and in the process shut down his central nervous system, or at least the part responsible for consciousness. Lester Brown lapsed into a deep coma.

Mr. Brown was 41 years old and had been using cocaine for six or seven years. Like all cocaine-related knockouts, Lester's occurred not from a steady accretion of drug but from a single, exciting snort. Within 30 minutes after inhaling the cocaine he was in our ED, comatose and intubated, his breathing fully supported by a ventilator.

A CT scan of his brain confirmed the clinical diagnosis. It showed blood everywhere, both within the ventricles of the brain and covering the outside surfaces. From CT scan he was sent back to the ED, where he awaited a bed in MICU [medical intensive care unit] to be opened up. While in the ED both neurosurgery and neurology consults were called. I also went down to the ED to evaluate him.

His brother, three years younger but equally large and sinister looking as Lester, gave us the history. 'Buba' Brown made no attempt to hide his and Lester's illicit drug activity; indeed, he recounted the details as if we knew all along about their habit and drug trade.

"We just did our thing, man, and this happened. We didn't try to hurt no one. We just dealt with dudes we didn't know too well. Shit, man, this could've happened to me. Mother fuckers!" He was plainly angry in a menacing sort of way but, for the moment, his anger was directed elsewhere. I scanned the ED and thankfully noted the two security guards on duty.

Although Lester's hemorrhage was explainable by a sudden whiff of pure coke – a well-documented cause of stroke – his brother believed in a more sinister cause.

"I told him not to buy off those guys," he said, as if I knew who "those

guys" were. "Lester, he didn't think he had enemies. Man, what are you going to do? I can't believe this happened to Lester. Mother fuckers! MOTHER fuckers!"

"What do you think they sold him?" I asked, more out of curiosity about the drug trade than belief that the answer would affect Lester's outcome.

"Never you mind," Buba said. "I know. And they know I know. Is he gonna make it? I mean, that's what I need to know now, Doc, is he gonna make it?"

"It's too early to tell but it doesn't look good. The neurosurgeon and neurologist are on their way to see him now."

* * *

It is axiomatic that all patients are treated without distinction in both the emergency department and intensive care units. The physical space, the level of nursing care, indeed the entire resources of acute care hospitals are the same for all ED and ICU patients. This *equal level* is not always true elsewhere in the hospital, where there can be distinctions between single vs. private rooms, private duty vs. staff nursing, care by private office-based physicians only vs. care by salaried staff physicians, etc. In the ED and ICU it doesn't matter if you are a pauper or king, drug addict or company president (or both). Care is delivered by the same people to everyone, and is based solely on assessment of the medical problems and the available resources.

Notwithstanding equality of care in these areas, most physicians consider drug addicts and dealers the lowest form of life. We accept that they will lie, cheat, steal and, if necessary, kill to get what they want. But in the ED and ICU even the lowest of the low receives the same top notch care as anyone else, even if their physical problem is 100% drug-related. Doctors and nurses often ponder the irony that the life we save today may rob (or harm) us tomorrow.

So Lester's drug habit and reason for coming to the hospital didn't matter. We would have done whatever necessary to salvage his brain regardless of the cause. Unfortunately there was just very little to offer. Within an hour of arrival to the hospital both neurosurgery and neurology consultants confirmed Lester's dismal prognosis.

Apart from the fact that the neurosurgeon operates, while the neurologist is mainly a diagnostician of nervous system disorders, a principal distinction between the two specialists is the amount of words they leave in a consultation note. The neurosurgeon always writes what is necessary in a

page or less. The neurologist seldom makes do with less than two pages, and sometimes writes four or five.

The neurosurgeon who consulted on Lester summarized the situation thus: "Massive SAH [subarachnoid hemorrhage, a hemorrhage into the brain]. Little hope for survival. Nothing to offer surgically." This assessment took him only about 10 minutes in the ED.

The neurologist's summary was a bit longer, and was not completed until after Lester had been transferred to MICU. He wrote: "In the presence of deep coma from SAH the prognosis for survival is very poor. Based on data from Levy, et al, (Annals Internal Medicine, March 1981, pages 293-301), patients like Mr. Brown have a 74% probability of dying from SAH. However, given his specific neurologic findings, particularly the absent oculocephalic response [eye movements when the head is turned] and absent corneal reflexes [blinking when the eyeball is lightly touched], Mr. Brown's chance for survival is only 5%, and even at that he would in all likelihood remain in a vegetative state. I simply cannot be optimistic because of the size of his bleed and level of coma. Suggest discussion with his family regarding DNR [do not resuscitate] status. Thank you. Will follow."

Eight vs. 117 words to say the same thing. Yet both consultants were helpful. The neurosurgeon told us that brain surgery was no use, and that Mr. Brown could be transported directly to MICU; a stop in the operating room was not a viable option.

The neurologist told us, authoritatively, what to expect and what to tell the family.

* * *

Once ensconced in MICU Lester had plenty of visitors. It was not easy to know who was related by blood and who by economic considerations. On his very first hospital day I was told he had "one brother and one sister." The next day, two brothers and two sisters came to visit. He had no wife but at least two "girlfriends." The only relatives I felt certain about were Buba, his younger brother, and their mother, an attractive, well-dressed woman in her early 60s.

To facilitate communication everyone agreed that Buba would be the official spokesperson, and I dealt only with him. Buba was seldom alone, however; at least a few friends and relatives usually stood nearby during our conversations.

As so often happens in cases where the principle affliction is self-induced, the patient's extended family did not easily accept the drug connection. Notwithstanding Buba's comments at the time of admission, I had the distinct impression that everyone else thought something amiss had happened to Lester *in* our emergency room. After all, they reasoned, Lester only came in because of a headache, albeit a severe one. "He wasn't that bad

when he left home," they commented. "He was always a strong man. Healthy as an ox." Buba did little to dispel this line of thinking, either because he could not make good on his promise of retribution or because he would not admit to dangers from snorting coke. I reassured them repeatedly that the care in the ED had been superb, adding that Lester might *not be here now* had the care been less than perfect.

As expected, Lester showed no signs of improvement. Despite full ventilator support, anti-hypertensive medication, and round-the-clock nursing care, he remained deeply comatose and unresponsive.

By Lester's third day in MICU his "family" had grown to twelve people, all sitting or standing vigil in the waiting room. Although only two people are allowed in a MICU patient's room at any one time there is no prohibiting an army, if it wishes, to camp out in the waiting area.

That afternoon I managed to catch Buba alone in Lester's room. I told him the outlook was dismal and that neither myself nor the neurologist thought Lester would survive another 24 hours. I quickly added that we would not stop the breathing machine or any other therapy, and that Lester would stay in MICU until he either died or got better. (No way was I going to even hint at a slackening of care, or ask for a DNR order, although by this point I felt intensive care was futile. Nonetheless it was important, to forestall any doubt about our hospital's management, that we continue with full support until the inevitable end.)

"Can you come out and explain to our family? They'll want to hear it from you."

I agreed and followed Buba out to the waiting room. The four or five relatives who were smoking (near a no-smoking sign) put away their cigarettes, and a woman on the phone hung up when she saw us coming. After everyone assembled in a corner of the large waiting area I began my explanation. I took the direct approach.

"Unfortunately there's been no improvement in Mr. Brown's condition. The neurology specialist saw him again this morning and repeated another brain wave test. It looks very bad. His brain activity is only one step above being what we call totally brain dead. At this point, in all honesty, we don't think he's going to make it."

"You're not giving up, Doc." A command, not a question, from one of Lester's relatives.

"Absolutely not," I retorted quickly. "I made that clear just now to Mr. Brown's brother and I promise all of you, we are in no way giving up. But Mr. Brown wanted me to tell you how it looks, and it looks real bad."

"Well, we're going to stay right here until he gets better. Did you find

out the cause yet?"

"We still think it's from cocaine," I answered matter-of-factly. "The CT scan of his brain, the cocaine found in his urine, and the physical exam all point to a ruptured blood vessel like we often see in cocaine users." I refrained from using the word addict.

"Sometimes this happens just from high blood pressure, but in Mr. Brown we're pretty certain it was the cocaine." I had gone over the likely chain of events several times already. This time I wanted to add, 'So all of you should quit using coke,' but didn't dare.

One of the young men in the crowd, a brother or friend or business partner, looked straight at me and said, menacingly: "It ain't no cocaine that did this." I didn't respond since his comment wasn't a question.

After answering a few more questions I returned to the ICU. I hoped I would not be on duty when Lester died, but at the same time felt ashamed at my wish; *somebody* would have to tell his family.

* * *

Lester's blood pressure collapsed the next afternoon. I was there. Futilely, we pumped on his chest and infused pressor drugs but it didn't matter. His brain was gone. We pronounced him dead at 4:35 p.m.

"Are you going to tell the family?" one of the nurses asked me. She was not asking *if* I were going to tell them, as if that was an option I could pass on. She was asking if *I* was going to take the responsibility or delegate it to someone else, such as a medical resident.

"Sure," I said. As director of the intensive care unit what else could I say?

I looked at the intern who had assisted with the resuscitation, an innocent fellow named Bob. It would have been unfair to send him out alone but he had to learn how to give out bad news. It was part of being a physician.

"Bob, why don't you come with me?"

We went out together. Walking toward the waiting area I mumbled to Bob that the only way to do this sort of thing is to be direct. He nodded in agreement. Lester's family were all very quiet as we walked toward them. I think they suspected that Lester had died and were just waiting to hear it from me.

"I'm sorry," I said. "Mr. Brown just passed away."

There were about two seconds of silence, then all hell broke loose. Lester's two girlfriends started wailing and sobbing. His mother began repeating, in a high pitched sing song, "Oh No! Not my Lester! Not my Lester!" She went on and on, literally a chant for the dead. Then the young man who knew "it ain't no cocaine" began pounding the wall with his fist, at which point Buba tried to restrain him. This action only backfired, as the

pounding man became more combative.

Somebody yelled at me, "Say it ain't so!" I feebly responded, with my head slightly bowed and in a manner to indicate that it was so, "I'm sorry."

Ordinarily I would stick around, answer questions, commiserate with the family, ask for autopsy permission. Not this time. Discretely, Bob and I retreated back toward MICU. I felt I had done my duty; they had been forewarned of Mr. Brown's eminent demise, and I had told them as soon as it happened.

On our way back we heard "*WHAM*!" and the sound of broken plaster. The young man had broken away from Buba and, incredibly, put his fist through the wall. The blow would have floored Muhammad Ali in his prime. Bob and I rushed inside MICU and called Security. I had a dead man inside and a crazy one outside. We could hear the wailing and crying and banging continue. I feared more for innocent bystanders than for myself or the MICU staff. In fact I felt protected in MICU; they wouldn't dare enter the sanctuary of the critically ill, where Lester now lay in repose.

Fortunately I was right. But outside MICU it was a different story. Less than a minute after my call two security guards appeared. Ordinarily two guards can take care of almost any hospital disturbance but in this case they were outnumbered. The young man's anger had infected the group so much that the guards could made no headway; reasoned discourse was to no avail.

Standard policy is to escort off the hospital grounds anyone causing a disturbance. But how do you escort a dozen wailing, fist-pounding, angry people who have just lost a loved one? Well, you can't. More security guards were called up, for a total of five.

A melee erupted. Punches were swung and jaws hit. Fortunately, no one had a gun or the melee would have added to our ICU census. As it turned out, one guard suffered a broken jawbone and one relative had his shoulder dislocated; both were treated in the emergency room.

I saw none of the action. For a full hour after the noise abated the MICU staff stayed put. Then our Chief of Security came in to report all was under control and that two officers would be stationed outside the ICU all night. Apparently one of the group had made some threatening remarks, necessitating the extra protection. There had been no arrests.

Nothing came of the threats and everything in MICU quickly returned to normal. Two hours after death was pronounced Lester's body was unceremoniously taken to the morgue.

Comment

Lester Brown was one of many cocaine abusers we treat every year. Famous victims – the college basketball star Len Bias, the professional football player Don Rogers, the movie star John Belushi – are the tip of an enormous iceberg of senseless, drug-related death. In any ED serving the inner city dozens of cocaine abusers are treated every week; on average, at least one of them will be ill enough to warrant hospitalization in intensive care. Hospital admission may be for a variety of cocaine-related problems: coma, cerebral hemorrhage, severe hypertension, cardiac arrhythmia, pulmonary edema, pneumonia. Most patients survive their first acute medical problem but many, like Lester, do not. Of course we never see the ones who die in the street, or in a crack house.

Sudden death from cocaine is usually related to either subarachnoid hemorrhage, heart attack (coronary thrombosis), or cardiac arrhythmia. The mechanism of SAH is poorly understood but is probably due to a sudden rise in cerebral blood pressure caused by the drug, followed by rupture of a vulnerable blood vessel within the brain.

Since the case of Lester Brown several other young people have died in our MICU after a single encounter with cocaine. Anyone who uses street cocaine risks sudden death.

-- END –

15. Crisis and Lysis

"Doctor Martin, I need some help." The call was from Bill Moody, one of our medical residents working in the emergency department.

"Sure Bill, what's up?"

"There's a patient down here with chest pain, shortness of breath and hypoxemia [low oxygen level]. He's in some distress and I think he might have a pulmonary embolism, or at least that's the only way I can put his story together. He and his wife just came back from Florida. They drove straight through, non-stop, eighteen hours in the car. I want to take him for a lung scan. But I wonder, should I heparinize him first?"

"Don't do anything yet, Bill. I'll be right down."

Well, I thought, here it is again. Pulmonary embolism. One of the most difficult diagnoses to make *and* treat. The great masquerader, undiagnosed in half the patients who have it. And often over-diagnosed in patients who end up having something else. Difficulty in making an accurate diagnosis of "PE" has long plagued physicians, but nowhere is the diagnosis more bothersome than in the emergency department. Make the diagnosis and the patient must be admitted to hospital. Rule out the diagnosis and, frequently, the patient can go home (maybe the chest pain was just indigestion). *Miss* the diagnosis and the patient can die.

The mere suspicion of "PE" is enough to engender anxiety in the medical staff. "I thought Mr. Jones might have PE" justifies ordering costly tests at any time of day or night. "I couldn't rule out PE in Mrs. Smith" explains why an otherwise rational physician might start a patient on dangerous blood thinners. And "I missed a PE in Mr. Harris" raises the specter of lawyers hunting you down for the inevitable lawsuit. Doctors who suspect PE often feel caught between the proverbial rock and hard place.

What exactly is PE? Embolus is from the Greek *embolos*, meaning "plug." If a blood clot forms in a leg vein and travels to the lungs it then becomes a pulmonary embolus (or embolism). Part of the blood circulation within the lungs captures the embolus and becomes, literally, "plugged" from the clot. If more than one blood clot travels to the lungs they are called pulmonary emboli.

Blood clots traveling to the lungs – pulmonary emboli – may arise from veins anywhere, but the legs are by far the most common site of origin. Until the clots break off and lodge in the lungs they are usually silent; only

infrequently do they cause any discomfort in the legs from where they came.

Once the traveling clots reach the lungs they can cause pain, shortness of breath, cough (either dry or with expectoration of blood), palpitations, sweating, and a host of other symptoms. The basic problem is that, given suspicion of a pulmonary embolus, there is no easy way to make the diagnosis, at least with certainty. Sometimes doctors have to treat the patient based on very inconclusive evidence. Standard therapy is intravenous heparin, a potent blood thinner tricky to use in the best of circumstances. Less frequently employed for PE is streptokinase, a drug more potent (and potentially more dangerous) than heparin.

If treatment was easy pulmonary embolism wouldn't be such a big headache for doctors. For example, patients with viral infections often receive antibiotics because the treatment is easy, even if it is likely to be ineffective. Unfortunately there is no simple treatment for PE. Because heparin 'thins' the blood by interfering with normal clotting, it puts the patient at constant risk of *bleeding.* Of course heparin also prevents more clots from forming and going to the lungs, but what's the good of that if the patient has a major hemorrhage?

Still, doctors use heparin because it stops existing clots in their tracks and keeps new ones from forming; the body's own defenses are relied on to break up the existing clots, which can take several days. Heparin is certainly adequate if the patient is not suffering shock or breathing difficulty, and can wait for natural defenses to eat away at the existing clots.

But what if the patient is in real distress because of the clots? Then streptokinase is indicated, since it acts directly to break up blood clots. But, because streptokinase is so potent (it dissolves clots good and bad), the patient may be at greater risk for bleeding than with heparin, for example after a surgical procedure. Also, streptokinase has not been shown to give better long term survival than heparin; survival with either drug is about three-fold over no treatment (which is why some treatment is mandatory if you make the diagnosis). Think of it this way: streptokinase heals the very sick patient quicker than does heparin, but the long term result (survival) is about the same.

In any case, the bleeding risks (with either drug) mandate reasonable certainty about the diagnosis. The major dilemma is over diagnosis, not the specific type of treatment (make the diagnosis and the patient will get treated with *something*). Often the choice is between the lesser of two evils: risk of bleeding from treatment vs. risk of more (and potentially fatal) clots traveling to the lungs.

Why is diagnosis so difficult? Because the symptoms are what doctors call non-specific, and also because there is no fool-proof diagnostic test. Chest pain and difficult breathing, two hallmarks of PE, are also common in heart attack, pneumonia, pleurisy, esophagitis, the flu, muscle spasms, and many other serious and not so serious conditions. Physicians can't treat

every chest complaint as a pulmonary embolus, but if the diagnosis is present and missed, that patient's *next* clot could be fatal.

Some situations increase the risk for pulmonary embolism: major surgery; prolonged bed rest or immobilization; pregnancy (particularly third trimester); chronic heart or lung disease; and cancer. Pulmonary embolism is rare in healthy, active people, with one major exception: women taking birth control pills, especially if they smoke.

Compared to pulmonary emboli the majority of heart and lung disorders – including pneumonia, heart attack, heart failure, emphysema, and lung cancer – are relatively easy to diagnose. By contrast, PE is both missed *and* over-diagnosed frequently. Trying to diagnose PE sometimes seems like trying to identify a close friend behind an opaque glass door. If you could open the door you would have no trouble; otherwise, you can only guess who is there.

The definitive diagnostic test involves passing a catheter through the heart and into the lungs, then squirting dye into the pulmonary blood vessels. This test, being invasive and expensive, is hardly routine. Instead, doctors in most hospitals rely on the lung scan, a nuclear medicine test that images the *effects* of the blood clots but not the clots themselves.

Unfortunately the lung scan is a very sensitive test; almost any lung condition can show up abnormal, including asthma, heart failure, emphysema, pneumonia, and so forth. Still, certain patterns on a lung scan favor the diagnosis of pulmonary emboli. Radiologists who interpret lung scans use terms like "high," "low" and "indeterminate" to describe the probability that a given lung scan represents pulmonary emboli and not something else.

Of course radiologists can only interpret images as they appear on film. The clinician has to incorporate the scan interpretation into his or her own *index of clinical suspicion*. *Clinical suspicion* is a catchall term for what a doctor suspects after examining and talking to the patient. Essentially the doctor caring for the patient asks: do I have a high or low clinical suspicion that my patient has a pulmonary embolus? And, given that index of suspicion, how do I use the results of the lung scan?

When all the rigmarole is dispensed with (including opinions from several lookers-on), it comes down to something like the outline shown below (PE = pulmonary embolism). It is seldom this simple, but the chart gives an idea of the diagnostic process. The problem is that "another test" is either the highly invasive catheter study, or some less invasive but usually less conclusive study, such as looking for leg vein clots that might (or might not) break off and go to the lungs.

Would Dr. Moody's patient be difficult-to-diagnose or straight-forward?

Scan reading: probability for PE	Clinical suspicion: for the diagnosis of PE	What to do for the patient
High	High	Treat for PE
High	Low	Do another test
Indeterminate	High	Treat, or do another test
Indeterminate	Low	Abandon the diagnosis of PE
Low	High	Do another test, or abandon the diagnosis
Low	Low	Abandon the diagnosis

* * *

In the emergency department I met Francis Jarvin, a 50-year-old stock broker just returned from Florida with his wife, Sylvia. I was struck immediately by two things: his breathing and his suntan. Each of his breaths was deep and painful, with the pain mostly felt over his right chest. In effect he 'splinted' his breathing, checking each inspiration to minimize pain made worse by chest movement; as a result his breaths were rapid and shallow. A normal respiratory rate is 10 to 16 effortless breaths each minute; his rate was 40. He wasn't shocky or confused and his vital signs were stable except for the rapid breathing.

He sported a loud Hawaii-type shirt, pink pants and white loafers, hardly the clothes we see in our Midwest patients (during March, no less). He was clean-shaven, with thinning hair and well manicured features, and sported a deep, even suntan and slight paunch. He could just as well be lying at the beach as on a hospital bed in our ED. The apprehension in his eyes, the too-rapid movement of his chest cage, and an oxygen mask that obscured his nose and mouth, told me he was in the right place.

About ten hours earlier, somewhere in Kentucky, Mr. Jarvin first noted chest pain and shortness of breath. He thought it might be indigestion and didn't want to stop in an unfamiliar city "if that's all it was." About an hour later his wife took over the driving. As soon as they arrived home she drove him to our ED. Given his level of discomfort I was amazed he'd been able to finish the car trip.

The oxygen mask and breathing problem made it difficult for him to talk. I needed to be quick and direct with my examination, but also try not to alarm him. I asked Dr. Moody to get me his chest x-ray, blood gas report, cardiogram, and emergency room chart.

"Hi, I'm Dr. Martin. I'll be taking care of you upstairs. I just want to

ask you a few questions, listen to your lungs and heart, and then you'll go for a lung scan. I understand you just came back from Florida." I spoke as if I was making conversation with an old acquaintance.

"Yes...that's right."

"I know you're short of breath and in some pain. Don't try to talk too much. Just answer yes or no if you can, or nod your head. You drove home by car?" He nodded yes.

"You drove straight through, all twelve-hundred miles, except for stopping a few times?" Another nod.

"What's the longest you sat in the car without stopping? Just given me an approximation."

"About...five...hours...I guess." Mr. Jarvin squirmed with chest discomfort.

"Did you ever have pain in your legs?"

He shook his head.

"I understand you're taking some pills for high blood pressure."

"...Yes, my wife has them."

"Calan and Dyazide, is that right?"

He nodded agreement.

"Have you ever been in the hospital before?"

He held up one finger.

"Once?"

"Yes...for hernia...about ten years ago."

"OK. I'm not going to ask you anything else right now. Just let me check a few things."

Over the next few minutes I examined his heart, lungs, abdomen and legs, reviewed his vital signs and initial lab results, and developed a firm impression: severe pulmonary embolism.

Dr. Moody came in just as I was finishing. "They're ready for him in nuclear medicine," he said. "Should we start him on heparin first?"

"No," I said. "He's going to MICU [medical intensive care unit] straight from the lung scan. Please call MICU and tell them we'll be there as soon as the scan is finished. Ask Marsha Ligner, the head nurse, to please order up streptokinase and have two hundred and fifty thousand units ready when we get there."

"You're going to use *streptokinase*?" Dr. Moody had not used this drug before.

"If his scan shows what I think it will, that's what he needs. Heparin isn't going to help him much right now. We need to break up these clots right away."

I accompanied Mr. Jarvin to nuclear medicine, located one floor below the emergency department, in the basement. I was nervous about him. What other diagnosis could this be? I didn't know, but we needed the scan to confirm pulmonary embolism and justify using streptokinase. I just hoped he would finish and get to MICU before some catastrophe occurred.

The technician injected his arm vein with radioactive technetium, the material used for showing up the lungs, then set the scanner over his chest. I watched the images as they appeared on the monitor. In healthy lungs the radioactive material is distributed evenly throughout the open blood vessels and shows up as a pattern of fine dots with no interruptions. Blood clots in the lungs disturb the distribution of radioactivity and cause blank spaces to appear on the scan. When examined along with the patient's chest x-ray, the particular pattern of blank spaces determines the probability for pulmonary embolism.

Mr. Jarvin's scan showed enormous blank spaces, pie-shaped defects in both lungs evident from any angle the camera scanned. When all the camera angles were completed I said, "let's go." I didn't need the radiologist to tell me what I already knew. His likelihood for embolism was "high-high."

In another three minutes we were in MICU. It was 2:30 in the afternoon.

"Marsha, do you have the streptokinase?"

"Ready to go."

"Let's hang it."

* * *

Mr. Jarvin needed both streptokinase and reassurance. As the drug began flowing into his arm vein, I explained the situation.

"You do have blood clots in your lungs," I said. "We've begun a drug that should help break them up. You'll probably continue to feel uncomfortable for a while, but you should feel better as the clots dissolve."

"I hope so," he winced.

And so did I. I thought back to Sabrina Johnson, 17 years old. So young. And I was younger too; it had been a decade since she showed up in our ED. A high school student, Sabrina presented to the ED with a "chest pains," or at least that's what her 34-year-old mother told the triage nurse. This was only three days after a previous hospital stay for therapeutic abortion.

I remember the girl well: a short, obese teenager, cherubic face. She spoke with a limited vocabulary and, perhaps being so young, did not seem particularly apprehensive about being back in the hospital so soon. She had a history of two pregnancies; the first one, just a year earlier, was brought to term. Her infant boy was being cared for by Sabrina's mother while she attended school.

Her mother related that Sabrina's pains were sharp and fleeting, and that's why she brought her back to us; in fact, the pains probably worried her mother more than Sabrina, who was claiming no pain when we examined her. Sabrina's legs were fat, too fat to properly examine for clots, but such exam is usually not useful anyway. Her oxygen level was normal, which it can be with pulmonary embolism. Her cardiogram and chest x-ray were, in the parlance of medicalspeak, "unremarkable," meaning not abnormal in any significant way.

I saw her in the emergency department right after she arrived, and discussed her case with the staff ED physician. What to do? We thought of pulmonary embolism, considered it a distinct possibility, and so ordered the requisite lung scan. But we didn't heparinize her right away (or even consider the use of streptokinase). She was not in any distress and actually seemed comfortable in the ED. Although "at risk" for PE (because of obesity and her recent pregnancy), it was just three days after her abortion and we didn't want to risk any bleeding unless certain of our diagnosis. Then too, Sabrina was only 17, had no chronic diseases, and her pains could just as well have been pleurisy, a benign viral syndrome. In looking back I would say our index of clinical suspicion was on the high side of low.

So we ordered the lung scan and while the technicians wheeled her to the basement, I stood around the ED to kibbutz with the staff. If Sabrina's lung scan was negative we would probably send her home; anything abnormal, and she would be admitted to hospital, and probably go on heparin therapy.

Ten minutes later we were paged 'stat' to the radiology department; Sabrina was now in distress, even before she had been injected for the lung scan. *Acute respiratory distress*: breathing labored, nostrils flaring, neck muscles contracting, unable to speak. Her pulse raced at 140 and worse, far worse, within a minute, as we stood there, she went into shock – NO BLOOD PRESSURE! From that point it was all downhill. Two highly trained ED doctors and myself did CPR for one hour and 10 minutes, right in the radiology department. We gave both heparin and streptokinase, plus assorted other drugs, but nothing worked. Sabrina was dead. And we knew the diagnosis, even without the scan. In fact, from the moment we saw her in distress we all knew.

The autopsy confirmed a massive "saddle" embolus straddling both main branches of her pulmonary artery, and completely occluding the flow of blood from her heart to her lungs. The pains she experienced before coming to the ED were most likely from smaller pulmonary emboli, sentinels sent out to warn of impending doom. Would she have survived if

we had treated her right away, in the ED? Possibly, but we'll never know. I do know that since Sabrina Johnson I am sensitized to this diagnosis, respectful of what massive pulmonary embolism can do and how quickly it can snuff out a life.

* * *

I had spoken to Mrs. Jarvin only briefly in the ED and during her husband's lung scan test. She deserved some explanation so I went out to the MICU lounge where she was waiting.

I guessed her to be about 48. Like her husband, she was also suntanned and dressed for Florida. She sported a bouffant hairdo anchored by heavy silver earrings, and wore pastel pants and a purple jacket. The clothes were flashy but her eyes suggested stress and fatigue. In dress and bearing she reminded me of sleep-deprived travelers stranded at an airport during a winter storm, all dressed up, incongruously, for warm weather. Events are totally out of their control, both the weather and decisions on when the planes will fly. All they can do is wait and hope and commiserate.

"Mrs. Jarvin," I said, "the scan shows massive blood clots, which explains all your husband's symptoms. We've just started the blood thinning medication, and he seems to be tolerating it well."

"This is serious, isn't it doctor. Can he die from this?"

I thought of Sabrina and almost blurted out 'You betcha!'

"That's always a possibility in this condition," I replied, "and that's one reason he's in MICU, so we can watch him closely. The next few hours will be crucial, but right now he's stable. Why didn't you stop at a hospital earlier, if he was having chest pains?"

"Why? Why?" she said mockingly. "Because he's bull-headed, that's why! I begged him to let me stop. He kept saying, 'we'll be home in a few hours, it's just indigestion. Keep driving.' So I kept driving. If he doesn't make it I'll never forgive myself."

"What exactly happened when you got home?"

"He could hardly walk. He certainly couldn't lift the suitcases out of the trunk. I thought he was going to collapse right on the driveway. He was sick, really sick. He could hardly catch his breath. It was all I could do to get him back in the car. I drove straight here. Didn't even go in the house. Too bad the kids are away." The Jarvins had two grown children living out of town.

"Well, you did the right thing, bringing him here as soon as possible, that's for sure."

Mrs. Jarvin stayed in the MICU lounge the rest of the afternoon and visited her husband a few minutes each hour. Their suitcases remained unpacked in the car.

In the emergency department Mr. Jarvin's oxygen saturation was 90%, a

low value considering that he was inhaling extra oxygen from the face mask; normal oxygen saturation without inhaling extra oxygen is over 95%. In patients like Mr. Jarvin we follow both their respiratory rate and oxygen saturation, the latter with a non-invasive 'pulse oximeter', a clothes pin-type apparatus that fits comfortably over the finger. A chronology of Mr. Jarvin's course that afternoon:

2:00 - Respiratory rate 40, oxygen saturation 90%
2:30 - Streptokinase begun
3:00 - RR 35, oxygen saturation 91%
4:00 - RR 31, oxygen saturation 92%
5:00 - RR 26, oxygen saturation 93%
6:00 - RR 24, oxygen saturation 94%

At 6:30 I found our patient comfortable, no longer in pain, actually smiling. His wife was in the room. "I feel much better," he said. "I don't know what you used, Doc, but it sure seems to be working."

I was glad we started streptokinase. Heparin would not have acted nearly so fast. If he didn't have a bleeding complication the prognosis was excellent.

Comment and Followup

Streptokinase was approved by the Food and Drug Administration in 1977 for treatment of pulmonary embolism, and has since been supplanted by newer fibrinolytic agents such as 'TPA'. These drugs have saved countless heart attack victims all over the world.

As a class, fibrinolytic agents are less commonly used than heparin in PE because, in some early studies, the drug seemed to cause more bleeding without improving survival. Yet they break up the clots quicker than heparin and are probably *not* any more dangerous. If used properly, and if the patient is not invaded frequently for blood drawing and other tests, the complications are about the same as with heparin. Certainly if we need to lyse a patient's blood clots quickly, then a fibrinolytic drug (such as TPA) is the way to go. Heparin is just too slow for that purpose.

We continued Mr. Jarvin's streptokinase infusion another 36 hours and then switched him over to intravenous heparin therapy. He stayed on heparin for several days and then began Coumadin, a blood thinner taken by mouth. There was no complication from any of the drugs and he went home on the eighth hospital day.

Mr. Jarvin took Coumadin for three months, by which time he was out

of the danger period. He never again subjected himself to prolonged sitting, the condition that led to venous stasis in his legs and formation of the clots.

Now he only flies to Florida.

-- END –

16. Extraordinary Care

Helga Bowman was 60 when her life went into a tailspin. Within one week of March, 1982, she learned of cancer in her right breast and emphysema in both lungs.

On March 12 she consulted a surgeon for a breast lump. Suspicious right away, Dr. Spivey explained it could be cancerous and that she might need a mastectomy. He biopsied the lump with a thin needle, removing just a few cells for microscopic examination. The next day, March 13, Dr. Spivey received a verbal pathology report: malignancy. He promptly called Mrs. Bowman, told her the diagnosis, and scheduled a mastectomy for March 16. Because of her long smoking history he also ordered pulmonary function tests and asked her to see me.

Mrs. Bowman came to my office on March 14. A middle-aged, kindly-appearing woman with a big smile, she displayed all the telltale signs of emphysema: raised shoulders; slightly pursed lips; slow gait; and contraction of neck muscles with each breath. Most emphysema patients are thin and she was no exception, weighing 110 pounds and standing five feet two. She wore a plain cotton dress and little makeup, and kept her brown hair brushed straight back. Any complaints? "I feel fine," she said, "like always. Just this lump I noticed last week." She pointed to her right breast.

"How long have you been smoking?"

"I started when I was about twenty. Forty years I guess."

"How much? Would you say a pack a day?"

"Yes, no more than that. But it never caused me any problems."

As she undressed for the exam I could see she was limited and less agile than other middle-aged woman with normal lungs. Her pattern of speech, an occasional pause to catch her breath, confirmed my visual assessment. She would look quite healthy in a family snapshot but not on videotape.

Her emphysema had developed insidiously. Shortness of breath on exertion and a chronic morning cough became such a routine experience for Mrs. Bowman that she considered the symptoms almost normal. "I thought they just happen when you get older," she explained.

Any lung capacity less than 50% of normal indicates marked breathing impairment; her capacity was only 42% of normal. This measurement, plus the chest x-ray and my examination, confirmed severe emphysema. Lung disease of this degree doesn't preclude breast surgery but does make one more cautious. Our concern is the general anesthesia, which can cause

problems for patients with respiratory impairment. I insisted that Mrs. Bowman quit smoking right away, and also prescribed inhalation treatments and corticosteroids for the two days before surgery.

She took the bad news and my recommendations surprisingly well. There was no denial, self-pity, or remorse sometimes seen in cancer patients. Just the opposite. The thing I remember most from her days before surgery was how *non-depressed* she seemed. Friendly and outgoing, she was the type of person who found something good in every bad situation.

On my visit to her hospital room the night before surgery I met Mr. Bowman, a 62-year-old retired fireman. Like his wife he was outgoing and affable. A short, thin man, he had the parched and wrinkled skin of an outdoors man. Fishing was his passion. "I've spent most of my life outdoors," he said.

"What do you catch around here?" I asked.

"Walleye. The lake's got tons of walleye this time of year. Just caught several last week."

We bantered a few minutes about the hazards of lake fishing and then I changed the subject to his wife's operation. I again discussed the risks of surgery and asked if they had any questions.

"No," they replied, with Mrs. Bowman adding, "We have faith in you and Dr. Spivey." Mr. Bowman nodded his agreement and that was that.

"By the way, you did quit smoking, didn't you?"

"Oh, yes," she said. "I'll never go back to cigarettes." The way she had accepted my no-smoking advice was a good sign. Whatever there was to do or take that might help, she would comply.

On March 16 Mrs. Bowman underwent a modified right radical mastectomy. The operation required removing her breast, some surrounding muscle tissue, and lymph nodes in the axillary (armpit) area that drain lymph from the breast. Several of these nodes were involved with cancer, indicating the tumor had spread outside her breast. She would need post-operative radiotherapy *and* chemotherapy to achieve any hope of cure.

She recovered from surgery without any problems and had an uneventful hospital course. Dr. Spivey discharged her a week later. In early April, as an outpatient, she began a regimen of 20 cobalt treatments, one each weekday. The x-ray beams were aimed to the region of her absent right breast and right shoulder area.

Two weeks later, after only 10 cobalt sessions, she developed a fever. Pus began to drain from her surgical wound. Radiation therapy was stopped and an antibiotic prescribed, but drainage from the chest wall continued. A few days later we re-admitted her to the hospital.

Skin bacteria had taken hold at the site of surgery and caused a deep, bony abscess in the underlying sternum, or breast bone. Despite heavy doses of intravenous antibiotics over several days the infection continued to spread, eroding almost completely through her sternum. Unchecked, the

infection might spread to her heart.

We called in a thoracic surgeon to debride the infection. The pain of deep debridement necessitated it be done under general anesthesia. Her lungs might not withstand another operation but there was little choice by this point. The infection had to be surgically removed.

At operation much of her sternum was like mush. The surgeon drained all the pus and removed several loose bone fragments, then sutured the remaining pieces of bone together with wire. The resulting assemblage of remaining sternal bone and wire did not form a solid breast plate. Although the operation effectively treated the infection, it left her with an unstable chest cage.

Mrs. Bowman was returned to MICU, still connected to the ventilator. Although her chest was covered with bandages I could see the problem. When she tried to breathe on her own, instead of the chest's normal outward movement with each breath, her chest collapsed *inward*. An unstable chest cage, added to severely weakened lungs, meant she would never be able to breathe unassisted by machine. The next day, April 10, she returned to the operating room for a tracheostomy.

Despite this major complication, and the reality that her cancer was far from cured, Mrs. Bowman remained cheerful. Metastatic breast cancer, severe emphysema, three operations, and several weeks in hospital did not daunt her spirit. She continued to have faith in her doctors and display a strong will to live, no matter what the setbacks.

At first I thought her upbeat mood was inappropriate, or at least an indication that she really didn't understand all that had happened. Certainly I would be depressed in her situation and so expected that she would be, if not clinically depressed, at least sullen, discouraged. Not her. This woman, who had two crippling diseases and a material net worth less than the yearly income of most doctors, viewed life only from the positive side. She had a good marriage, healthy children and grandchildren, and everything to live for. Why be depressed?

I did not tell her that getting off the ventilator seemed unlikely (I did tell Mr. Bowman). Instead, I accented the positive. "We're going to start chemotherapy. We can't give you any more radiotherapy but the drugs should help control your tumor."

"Good," she said with her lips. The tracheostomy tube prevented speech but her facial expression said, 'let's get on with it so I can get better and go home'.

Over the next month we gave her the latest in cancer chemotherapy. She developed complications, first pneumonia, then urinary tract infection,

then a stormy course of high fever and sepsis. At one point we thought she would not survive, but her body rallied and the infections were brought under control. She remained cheerful. The same could not be said for her doctors.

The oncologist and radiotherapist decided that she could receive no more cancer therapy and signed off her case. There was no evidence of tumor recurrence, but her fragile state made it impossible to give additional treatments if the tumor did recur.

At the end of a month in MICU, after her recovery from sepsis, I again tried to wean her off the ventilator. As expected, the effort was unsuccessful. Her lungs and chest wall were all but destroyed. Thus it came to be that, two months after the initial diagnosis of breast cancer, Mrs. Bowman lay in a MICU bed as a pure "pulmonary" case – and a disposition dilemma. Where could we send a 60-year-old, ventilator-dependent woman?

Ordinarily we would have looked for one of the rare nursing homes that accept ventilator patients. She would not consider it. Neither she nor her husband wanted her to be in a nursing home. Mr. Bowman had the solution.

"I'll take her home with me."

Before Mrs. Bowman's case, all of Mt. Sinai Medical Center's ventilator-dependent patients either died in the hospital or went to a nursing home. I knew that other hospitals had sent one or two ventilator-dependent cases home, but those patients were wealthy. The Bowmans barely made ends meet on Mr. Bowman's modest pension.

Artificial ventilation at home requires an oxygen supply company able to manage the ventilator and someone to care for the patient round the clock. The first requirement is easy, since third party payers will cover the ventilator care at home (it is cheaper than in a hospital). The second requirement is the usual stumbling block. Continuous nursing care is prohibitively expensive and insurance companies will not pay for it. Family members can theoretically do the job, but who has family that can dedicate to such extraordinary care? As it turned out, Mrs. Bowman did: Mr. Bowman.

I had my doubts. Mr. Bowman was devoted to his wife, but could he alone do the tasks that, in the hospital, require several shifts of workers? We would not discharge Mrs. Bowman until she was stable, but even then she would require almost constant attention. She needed to be suctioned often, given a bed pan when necessary, and fed three meals a day; the ventilator had to be checked instantly if any alarms went off; and calls had to be made if something went wrong.

Yes, yes, yes, Mr. Bowman insisted. He could do whatever was needed. All we had to do was show him what and how. Having no reasonable alternative, we agreed to send Mrs. Bowman home with a ventilator.

It took about a week for the oxygen supply company to set up a ventilator in their home. To make things easier we insisted on the same

model used in the hospital. The company first surveyed the house and decided the only place for the machine was a tiny first floor den. It could not go into their larger second floor bedroom because the connecting oxygen tanks were too heavy for any area but the basement. To properly connect the ventilator to the oxygen tanks required that the machine be located on the first floor. (Hospitals use a built-in liquid oxygen system with outlets in every patient room.)

After two weeks of discharge planning, during which Mr. Bowman received instruction from therapists and nurses, Mrs. Bowman was ready to leave the hospital. On the trip from MICU to the emergency room, where the ambulance was waiting to transport her home, a respiratory therapist breathed her lungs manually, with an AMBU bag. In the ambulance she received ventilation from a portable, battery-operated ventilator. Besides the two ambulance attendants, a nurse and respiratory therapist also came with her. Mr. Bowman followed the ambulance in his car and I followed in my car. I also took along our head of Respiratory Therapy; this was a first for him as well.

I had never been to her neighborhood and knew only that it was in a blue collar district about five miles south of the hospital. We followed Mr. Bowman off an expressway exit that I had passed many times before. Less than 500 feet from the exit the ambulance stopped in front of an aging, two-story wood frame house. Across the street was a gas station and a bar.

She lived on the fringe of a poor working-class section of town, in a house that was modest even by neighborhood standards. On the lot next door a rusting 1960s Ford rested on cinder blocks, its wheels and one door missing. Adjacent to the gas station was a vacant lot packed with half a dozen partly-stripped cars. In the air was the din of expressway with its high-decibel trucks roaring by every few minutes, and also a pervasive smell of exhaust fumes. Though not a slum, the neighborhood was far from appealing. I looked around and thought: *this* is where we are sending our first home-ventilator patient?

As for the Bowmans, they were glad to be home. Mr. Bowman made no apologies for the place, either direct or implied. "Come on in," he welcomed with enthusiasm.

The attendants took Mrs. Bowman out of the ambulance on a stretcher and, while the nurse bagged her manually, carried her up two concrete steps and into the house. Mr. Bowman directed everyone to her new bedroom, a linoleum-floored space approximately 12 feet square. Its walls were covered with musty, stained paneling and a single window looked out on the freeway exit (in the distance I could see a large green overhead sign pointing the way

to 'Downtown'). In one corner of the room was her bed, really a cot. Next to the cot stood a Puritan Bennett model MA-1 volume ventilator, its hoses arched in the air, ready for connection. What an incongruous sight!

We moved Mrs. Bowman from the stretcher to the bed and connected her to the ventilator. Mr. Bowman fussed with her a little, fixing her pillow and a blanket, while we checked her tracheostomy tube and then connected the ventilator. I turned the machine on. Whoosh! Whoosh! It worked fine. All the alarms were checked. While the therapists gave Mr. Bowman a final review of the machine's functions I briefly examined our patient. Her vital signs were OK and she was comfortable in her 'new' quarters. Careful planning had made the transition go quite smoothly.

The plan was to have the oxygen company's respiratory therapists visit the house twice a week, and for them to call me with any medical questions or special problems. Mr. Bowman planned to be home all the time; he would call for help as needed. On occasion their daughter might be able to help out, but responsibility for round-the-clock care clearly rested with Mr. Bowman.

Before leaving the house I walked around to see the other rooms. All were small, musty, and worn. The dwelling was not dirty; the kitchen, in particular, seemed clean, but I remember feeling depressed about the place and wondering why. Every room, apart from being tiny and cluttered (an ironing board, old TV, fishing equipment, and cardboard boxes filled with magazines occupied half the living room), had that worn-out-linoleum look. No matter how hard you cleaned this house it would always seem tired and old. Was this place a proper abode for a ventilator-dependent patient?

A half hour after we arrived Mrs. Bowman was safely in her bed and stabilized. After making sure everything checked out we said goodby and left. Outside, I looked at our chief therapist and he looked at me. We each had the same thought.

"Not very comfortable quarters, is it?" he said.

"No, her room at the hospital was bigger."

"Well, her husband's really devoted. He's learned everything we can teach him about the ventilator."

"It's amazing," I said. "You hear some homeowners complain about their crabgrass, or problems with the swimming pool, or their need for a hot water tub. The Bowmans don't have a pot to piss in and he brings her home on a ventilator!"

The external appearance of the house, the freeway-exit location, the size of the rooms, and the interior decor all made for a very depressing situation. But Mr. Bowman was not depressed, nor was his wife, the woman with cancer and emphysema. *We* were depressed. How long could she live under these circumstances? In this house?

* * *

During two months in the hospital, almost all of which was spent connected to a ventilator, Mrs. Bowman had suffered innumerable invasions of her artery for a sample of blood. We needed the blood to check oxygen and carbon dioxide levels and thereby adjust ventilator settings. I had never cared for any viable patient on a ventilator without obtaining at least one arterial blood sample a day.

We had also ordered numerous chest x-rays (once a day while she was in MICU) and many other tests, even when she was stable and only waiting to be discharged.

I did not know how often we would do *any* tests now that she was home. I didn't even know when I would get to see her again. The oxygen company took good care of the machine, and their technician called me once a week to report on her progress. "She's doing well," he said. "No problems with the ventilator. Mr. Bowman is managing things better than we expected. We just go in and do the maintenance checks." Given her benign course, I saw no compelling reason to make a house visit anytime soon.

About a month after she left the hospital I got a call from her husband. "Her tracheostomy cuff is leaking. I think it needs to be changed." The cuff is a small inflatable balloon at the end of the tracheostomy tube; it is normally inflated with air to provide a seal inside the trachea. If the cuff leaks and deflates, the ventilator cannot deliver the proper amount of air to the lungs.

"Is she getting enough air?" I asked.

"Yes, right now she is. But I have to keep inflating the cuff about every hour or the ventilator alarm goes off." Changing her tracheostomy tube was the one thing Mr. Bowman did not feel comfortable doing. It required disconnecting the ventilator for a minute, and that made him anxious.

"OK. I'll be out this afternoon," I said, and made plans to stop on my way home. Her house was out of the way but I didn't mind. I welcomed the opportunity to help Mr. Bowman, even with a small service. I arrived about 6 p.m. and found that her tracheostomy cuff was indeed leaking. I had brought two spares and used one as a replacement.

Mrs. Bowman was all smiles when she saw me. After I changed the tube she wrote on her pad: "Thank you."

"Are you having any problems?" I asked. "Anything bothering you? "No," she wrote. "I feel good." Examination showed that she was about the same as when she left the hospital.

Outside in the yard, where Mr. Bowman preferred to talk, I asked him, "How are you managing?"

"Just fine. I sure am glad you came out. That's real nice of you."

"No problem," I said. "Glad to do it. She seems to be holding up pretty well. Have you been out of the house since we brought her home?"

"No, our daughter does the shopping and I just stay here and take care of Helga."

"Can you do this all by yourself?"

"Sure. Maggie [their daughter] helps with the cooking sometimes, but I'm a darn good cook too. We do OK."

'Amazing', I thought, this man deserves some kind of medal.

Two months went by before I got another call. Same problem. Well, I thought, that seems about right. These trach cuffs last only about eight weeks. I went out to the house and changed her tracheostomy tube.

By my exam she seemed to be doing well. There were no bedsores that you often see in nursing home patients and her spirit remained remarkably good. Mr. Bowman was taking good care of his wife. I had thought of bringing a syringe and drawing some blood samples but decided against it. Four months had passed and nothing untoward had happened. What would I do with the results?

Outside I asked Mr. Bowman, "Have you been able to get away yet?"

"I've been to the store a couple of times. Maggie stays with her then, but its only for a short time."

"Can't you get away more often?" I was concerned about him.

"No. Maggie's got two kids of her own to care for. I'm doing fine," he said. "Don't worry about me. I don't mind staying home."

I decided not to push it.

* * *

Over the next two years I made another eight house calls, all to change her tracheostomy tube. I always found her cheerful and appreciative of my visits. She was never out of bed when I came, although I knew she did sit in a chair on those occasions when Mr. Bowman lifted her out. As for her husband, except for an occasional foray to the store he did not leave the house. He had not gone fishing since we brought Mrs. Bowman home from the hospital.

During those first two years Mrs. Bowman did not have a single blood test or chest x-ray. Word began to spread through the hospital of this indomitable woman at home on the ventilator, in whom not a single blood gas had been drawn in two years. I gave all the credit to the patient and her husband.

One day in August 1984 Mr. Bowman called. "She's got some pain and can't see out of one eye," he said. She had become suddenly blind in her right eye and his description made me suspect a detached retina.

"Call the ambulance," I said. "She needs to be seen by an

ophthalmologist."

Two years and two months after leaving Mt. Sinai Hospital she came back to our MICU. An eye surgeon saw her right away, diagnosed a detached retina and recommended surgery the next morning. It would have to be done under general anesthesia.

To prepare Mrs. Bowman for eye surgery I did an arterial blood gas and other routine tests. All the results were normal or unchanged from 1982. The operation was a success and she went home a week later, her vision improved.

* * *

I followed Mrs. Bowman at home for three more years. During this period I changed her tracheostomy tube about every two to four months. (I always waited for Mr. Bowman's call and always came the same day. I was out of town once and then my partner went to the house.) In those three years she had no blood tests or chest x-rays.

One day in June 1987 Mr. Bowman called. "Doc, she's not doing well. Her feet and belly are swollen and she's falling asleep all the time."

"How long has this been going on?"

"Oh, about a week."

"Does she complain about anything? Any pain or difficulty breathing?"

"No, she just looks bad, Doc, real bad."

"I'll be right out."

I feared the worst, but took some diuretic and heart medicine with me in case her problem was a simple case of congestive heart failure. Maybe I could treat her at home and keep her more comfortable than she would be in a sterile hospital bed.

Pulling up to the house I mentally noted the scene: not much different from five years earlier, except that the lot filled with junk cars was now vacant and the Ford on cinder blocks was gone. The expressway hummed as before and the Bowman's house appeared about five years more decrepit.

Mr. Bowman greeted me just as I got out of the car. He repeated his earlier observation. "Thanks for coming, Doc. She looks bad, real bad."

I went to the room and found her very lethargic. I shook her gently and raised my voice: "Mrs. Bowman! Mrs. Bowman!" She smiled and acknowledged my presence but the old cheeriness was no longer there. Both feet were engorged with edema fluid but I quickly discerned a different reason for the abdominal swelling. Her liver was rock hard, the first indication after five years that the breast cancer had spread. I went outside to

talk to Mr. Bowman.

"I'm suspicious that her cancer has come back," I said.

"I was afraid of that."

"There's little we can do if that's the case. She's had all the therapy possible and there's nothing else to offer."

"Can she go to the hospital and die there? The grandchildren are always coming over and..." He started to cry. I fought the same urge.

"Sure," I said. "Just wait here. I'll take care of it. Let me make a phone call."

I went to the kitchen to use the phone. After making the arrangements I returned to Mr. Bowman, who was still outside the house.

"The ambulance will be here in a few minutes. I've also phoned the hospital. We'll have a bed for her in the intensive care unit."

"Thanks, Doc." He was still crying.

Follow-up

Our tests confirmed that the breast cancer had recurred and spread to her liver. We kept Mrs. Bowman in MICU for two days and then transferred her to a regular ward, where she remained connected to the ventilator and received non-intensive nursing care. We continued intravenous fluids and kept her comfortable with morphine injections. She died in her sleep a week later.

Mr. Bowman accepted the loss. He was grateful for their last years together. After a five year hiatus, he returned to fishing regularly for walleye. And he still lives in the same house, alone, although his grandchildren visit often.

-- END –

17. Thyroid Storm

I'll never forget the patient Roberta Smith, a 35-year-old mother of two who presented to our emergency department with fever, confusion and a very fast heart beat. After a brief evaluation in the ED she was sent to MICU.

As they rolled Mrs. Smith into MICU on a stretcher I could *see* her problem. Set inside a thin face of smooth and shiny complexion were eyes that bulged like a frog's. A large mass straddled her windpipe and neck veins on either side pulsated rapidly with the rhythm of her heart. She

manifested the classic picture of an extremely overactive thyroid gland, so extreme as to deserve the appellation 'thyroid storm.'

I examined the ED ledger and x-rays while the nurses put her to bed. The admitting diagnosis was actually pneumonia, with "possible hyperthyroidism" a secondary diagnosis. On chest x-ray her pneumonia showed up as a hazy infiltrate in one lung.

There is no test comparable to an x-ray for diagnosing hyper-thyroidism, which is why the ED doctor hedged his diagnosis. Laboratory confirmation is based on blood tests that take much longer to process than any x-ray. Since hyperthyroidism is rarely life-threatening, doctors always wait for blood results if the patient is not acutely ill. In cases where the diagnosis seems apparent *and* the patient is toxic, treatment is begun immediately.

I asked Mrs. Smith a few questions but she did not answer. Instead she just stared at me with eyes bursting out of their sockets and moaned: "*Ummmmmmmmmmmmmmmmmm. Ummmmmmmmmmmm.*"

How had this woman gone untreated for so long?

* * *

The thyroid gland straddles the trachea or windpipe. Although the gland is normally not visible, it can be felt in some people as a slight bulge in the neck area on either side of the windpipe. Mrs. Smith's thyroid gland was at least triple normal size and easy to see. Size alone doesn't indicate over activity; some of the largest thyroid glands, or goiters as they are called, are inactive. Mrs. Smith's gland was both big and overactive.

Thyroid hormone regulates the body's metabolism. When there is too little hormone the patient is said to be 'hypothyroid.' Hypothyroid patients frequently complain of feeling sluggish and fatigued. They are also intolerant of cold temperatures easily tolerated by people with a normal thyroid gland. Hyperthyroid patients on the other hand – those with too much thyroid hormone – are often nervous, jittery and tachycardic. The most extreme cases of thyroid imbalance, high or low, represent a medical emergency.

The clinical exam of Mrs. Smith reads like a textbook case of hyperthyroidism:

- Skin warm and moist.
- Temperature 102 degrees.
- Pounding heart, rate 150 per minute.
- Wide, bulging eyes showing twice as much white sclera as normal

(medical term: exophthalmos)

- Large thyroid gland
- Muscle weakness in all extremities
- Muscle reflexes 4+ (hyperactive)
- Fine tremor in both hands
- Intolerant of heat (does not keep bed covers on)
- Displays emotional lability, ranging in a span of only 20 minutes from unresponsive and moaning to smiling euphoria to crying to laying quiet.

Mrs. Smith's medical diagnosis was Graves' disease, a disorder of altered immunology in which the thyroid's production of hormone is unchecked by normal feedback mechanisms. As is often the case with acutely thyrotoxic patients, she had probably suffered 'smoldering hyperthyroidism' from Graves' disease for many months. Pneumonia then pushed her over the brink and into a state of severe thyrotoxicosis (thyroid storm).

(In the spring of 1991 President Bush developed a cardiac arrhythmia – atrial fibrillation – from previously undiagnosed Graves' disease. Two years earlier Mrs. Bush also was diagnosed with Graves' disease. Familial association, i.e. Graves' disease among blood relatives, has been long known but occurrence in spouses is rare; as far as anyone knows the disease is not 'catching' or communicable. In any event their hyperthyroidism was a much milder form than that seen in thyroid storm.)

* * *

Robert James Graves was an Irish physician who lived from 1795 to 1853. He is known mainly for his classic paper on hyperthyroidism, published in the London Medical and Surgical Journal in 1835; a portion of this paper is quoted below. (From Ralph H. Major, M.D., *Classic Descriptions of Disease*, 3rd Edition, 1945. Courtesy of Charles C. Thomas, Publisher, Springfield, Illinois.)

NEWLY OBSERVED AFFECTION OF THE THYROID GLAND IN FEMALES

I have lately seen three cases of violent and long continued palpitations in females, in each of which the sample peculiarity presented itself, viz. enlargement of the thyroid gland, at all times considerably greater than natural, was subject to remarkable variations in every one of these patients. When the palpitations were violent the gland used notably to swell and become distended, having all the appearance of being increased in size in consequence of an interstitial and sudden effusion of fluid into its

substance...

A lady, aged twenty, became affected with some symptoms, which were supposed to be hysterical. This occurred more than two years ago; her health previously had been good. After she had been in this nervous state about three months it was observed that her pulse had become singularly rapid. This rapidity existed without any apparent cause and was constant, the pulse being never under 120, and often much higher. She next complained of weakness on exertion, and began to look pale and thin. Thus she continued for a year, but during this time she manifestly lost ground on the whole, the rapidity of the heart's action having never ceased. It was now observed that the eyes assumed a singular appearance, for the eyeballs were apparently enlarged, so that when she slept or tried to shut her eyes the lids were incapable of closing. When the eyes were open, the white sclerotic could be seen, to a breadth of several lines, all around the cornea...

[

(Since the disease is named after Robert Graves, the grammatically correct spelling should be Graves's disease. However, almost all modern medical texts, as well as unabridged dictionaries, spell it Graves' disease.

Graves' disease is an "autoimmune" phenomenon, which means the patient's own antibodies attack some portion of the thyroid gland, altering the normal regulation of thyroid hormone production. This may be the mechanism but it does not explain the basic cause (why do some people generate antibodies to their own thyroid?), which remains unknown. Stress was once thought to play a role but this has never been substantiated.

Graves' disease afflicts an estimated one million Americans every year, with women contracting the disease much more commonly than men. Usually the patient with Graves' hyperthyroidism has a benign course and can be treated as an *outpatient.* What made Mrs. Smith's hyperthyroidism so special was its *rapid and severe* presentation.)

* * *

The account by Graves was soon followed by that of Carl A. von Basedow, a German contemporary. Basedow's article appeared in a German medical journal March 28, 1840, part of which is quoted below. (From Ralph H. Major, M.D., *Classic Descriptions of Disease*, 3rd Edition, 1945. Courtesy of Charles C. Thomas, Publisher, Springfield, Illinois.)

Madame F., brunette, well built, of a decided phlegmatic temperament [married and had four children]. Madame F. felt herself very exhausted, suffered from an obstinate diarrhoea, had night sweats, lost a great deal of weight; at which time the eyeballs began to protrude from the *Orbita*. The patient complained of shortness of breath; she had a very rapid, small pulse; a resounding heart beat, she could not hold her hand still, spoke with a striking rapidity; and she liked to seat herself (because she always felt burning hot) with naked breasts and arms, in a cold draft. She showed unnatural excitement and carelessness about her condition. She went around a great deal, without being at all disturbed about her striking appearance in company. She satisfied without any afterthoughts her various strong appetites, slept well, however with open eyes.

Sometime in 1837 however, all of these symptoms increased in intensity...in the neck there appeared a strumous swelling of the thyroid gland; the area of pulsation of the heart was now broadened, pointing to enlargement ...the hastiness of speech and the unnatural excitement of the patient still more increased, night sweats, very offensive; urine scanty and read and, considering the continued diarrhoea, the appetite was always too strong. As far as the eyes were concerned, they were pushed out so far that one could see below and above the Cornea...the eyelids were pushed wide from one another; could not be closed with every effort. The patient slept with eyes entirely open.

...For a long time, the rumor was widespread in our town that this patient was crazy and was soon going to be taken to an asylum, and in fact she had an unfriendly attitude towards the physician; she never had, however, and that I can assure you, any insane ideas; she never showed any abnormal desires and if her astonishing carelessness over her truly sad condition seemed to be the result of her phlegmatic temperament, so the hastiness of her speech, the uncertain holding of her body and her hands, the tendency to go about naked or very lightly dressed, were undoubtedly symptoms of her heart disease.

Graves did not describe any treatment for his three patients. Basedow, for his well-built brunette, first administered leeches, then "spring waters" with "marked improvement." Based on modern understanding of hyperthyroidism, *no* 19th century therapy could have been expected to benefit the patient.

For decades after their original accounts hyperthyroidism was known as *both* Graves' and Basedow's disease. The preeminent clinician of the late

19th and early 20th century in this country was William Osler, one of the founders of Johns Hopkins medical school. In the first edition of his comprehensive textbook of medicine, published in 1892, Osler wrote:

> **EXOPHTHALMIC GOITRE (GRAVES'S DISEASE; BASEDOW'S DISEASE).**
>
> *Definition*. A disease of unknown origin, characterized by exophthalmos, enlargement of the thyroid, and functional disturbance of the vascular system...The disease is rare in men. Worry, fright, and depressing emotions preceded the development of the disease in a number of cases.
>
> *Symptoms*. In the acute form the disease may develop with great rapidity. In a patient of J.H. Lloyd's, of Philadelphia, a woman, aged thirty-nine, who had been considered perfectly healthy, but whose friends had noticed that for some time her eyes looked rather prominent, was suddenly seized with intensive vomiting and diarrhoea, rapid action of the heart and great throbbing of the arteries. The eyes were prominent and staring and the thyroid gland was found much enlarged and soft. The gastrointestinal symptoms continued, the pulse became more rapid, the vomiting was incessant, and the patient died on the third day of the illness.
>
> *Treatment*. Medicinal measures are notoriously uncertain... Treatment of the thyroid gland itself is rarely successful, and the operative measures have not been very satisfactory.

In the United States today the disease described by Osler is known only as Graves' disease. In Europe 'Basedow's disease' is the preferred eponym.

The first effective treatment for any cause of hyperthyroidism was surgical removal of the gland, an operation called thyroidectomy. Although it was not very successful in Osler's time, by the 1920s, with better control of infection and bleeding, thyroidectomy was accepted treatment for large goiters, including those associated with hyperthyroidism.

The first effective drug therapy was inorganic iodine, a method first reported in 1923. In small amounts inorganic iodine stimulates thyroid production but in large amounts it causes the opposite effect – a lowering of thyroid hormone. Initially, inorganic iodine was used only to prepare patients for thyroidectomy; the operation is less risky if the patient's thyroid

gland is first put to rest.

In the 1940s both specific anti-thyroid drugs and radioactive iodine were introduced (RAI), making it possible to treat hyper-thyroidism *without* surgery. One of the specific anti-thyroid drugs, propylthiouracil or PTU, is still widely used today.

Although PTU and inorganic iodine block overactive thyroid function, they are not definitive treatment. If the patient stops the medication hyperthyroid symptoms usually recur. To permanently treat hyperthyroidism the gland must be ablated. Ablation can be accomplished either surgically or by radioactive iodine (RAI).

RAI comes as a liquid preparation that is swallowed by the patient. The radioactive iodine enters the blood and is taken up by the thyroid gland, where it destroys active thyroid cells. As a result, the gland shrinks in size. Most patients who take RAI ultimately become *hypo*thyroid within a few years. Fortunately *hypo*thyroidism is easier to treat than *hyper*thyroidism, so RAI remains an accepted and well tolerated therapy.

In summary, there are three different methods for treating hyperthyroidism: long-term anti-thyroid medication (such as PTU); RAI (a liquid that is swallowed once); and surgery. Each therapy has good and bad features. Generally, radioactive iodine (RAI) is avoided in women of child-bearing years. Surgery is preferred if the patient is non-compliant with taking medication. In middle-aged, compliant patients, either RAI or anti-thyroid medication is preferred. (Both President and Mrs. Bush received RAI therapy.)

Ultimately the decision as to which type of therapy is based on the patient's particular circumstances and the experience of the treating physician.

* * *

I reviewed Mrs. Smith's case with Dr. Joel Stanley, one of the MICU interns.

"Joel, have you ever seen this before?" I asked, referring to her thyroid disease.

"Graves' disease? I've seen Graves' disease, but nothing like her."

"They started penicillin in the ED," I noted. "That's probably OK for her pneumonia. They also drew all the necessary thyroid function tests. We'll get those results in a couple of days but we have to treat the hyperthyroidism now. What do you want to give her?"

He pulled out his spiral-bound book of intensive care therapy. "Let's see. They recommend several possible treatments."

"By the time you read that book she could be much worse," I replied in a friendly manner. He was certainly right to research the proper therapy but she needed treatment right away.

"Joel, while you're reading, let's give her one milligram of intravenous propranolol."

"They mention propranolol," he said, pointing to a paragraph in his book.

"Good. I hope so. It's probably the best thing to use initially in thyroid storm. Propranolol doesn't treat the hyperthyroid gland directly, but it'll slow down her heart. Then we can work on the thyroid gland itself.

Mrs. Smith's heart was in danger of failing from too rapid a rate. Propranolol, a popular cardiac drug also known as Inderal, is excellent for modulating an extremely fast heart beat.

"What do you want to give next?" I asked him.

"The next step should be an anti-thyroid drug."

"Right. Let's start propylthiouracil, two hundred milligrams every four hours. We'll have to put it down a nasogastric tube. I don't think she can take anything by mouth right now. What else besides PTU?"

"Well," he said, "I suppose we ought to add some inorganic iodine."

"Good idea," I said. "Let's start potassium iodide. Put five drops in some orange juice and put it down her NG tube every six hours."

Iodine is part of thyroid hormone. Giving the drug in its inorganic form helps block thyroid hormone release from the gland. Since inorganic iodine works more quickly than any other anti-thyroid drug we usually give it to the sickest patients, starting about one hour after the PTU.

After we began Mrs. Smith's treatment I went out to look for her family. No one was around. Apparently her husband had left the emergency department as soon as she was transferred to MICU. A call to the phone number listed on the ED sheet went unanswered.

Two hours later Mr. Smith showed up at the hospital again. I explained his wife's condition, and told him she was receiving treatment for both pneumonia and hyperthyroidism.

"How did she get like this?" I asked.

"What do you mean?" he asked innocently.

"Well, hyperthyroidism rarely comes on all of a sudden, without any warning. How long has she been so sick?"

"I don't know," he confessed. "She hasn't been feeling well for a while. She's had some diarrhea and headaches for about a week, but we just thought it was the flu. She made an appointment to see a doctor, but that was for next week. Then last night she started coughing and running high fever. She felt no better this morning so I brought her here right away."

"Have you noticed her eyes getting more prominent?" I asked. "Well, yes, now that you mention it. But only in the last couple of weeks."

"So what happened today?"

"This morning she couldn't get out of bed to help our older kid get ready for school. She said she didn't feel well. I got him off to school and then went to check on her. She started acting like she didn't know me, and that's when I noticed that funny stare in her eyes. I got scared and just picked her up and brought her here."

"What did the doctors say downstairs?"

"They talked to me for a few minutes and said her problem was pneumonia and maybe an overactive thyroid gland, just like you said, and that she had to go to intensive care. They said she'd be OK but she had to be watched very closely. I still had the baby with me, so I left to take her to the sitter's, where she is now. Then I came back."

"Has your wife ever had a thyroid problem before?"

"No, not as far as I know. But she has been kind of high strung for the past year. She saw a doctor about six months ago and he gave her some Valium, but we never knew it might be from this thyroid gland. At least he never said anything about it."

* * *

By the next morning Mrs. Smith was much improved. Her pulse was down to 110 a minute and she was coherent. A follow up chest x-ray showed clearing of her pneumonia, so penicillin seemed to be the right antibiotic. In another 24 hours she was able to leave MICU.

Follow up

Mrs. Smith's pneumonia continued to improve and she stayed in the hospital only three more days. Blood tests begun on the day of admission confirmed a markedly over-active thyroid gland. Potassium iodide was discontinued at the time of discharge. For the next several months she took only PTU for her thyroid disorder.

Despite PTU she continued to experience intermittent symptoms from hyperthyroidism. After six months of PTU her endocrinologist decided to administer radioactive iodine (she planned to have no more children). As expected, the RAI obliterated most of her thyroid gland and she became *hypo*thyroid several months later. Thyroid hormone replacement was started as soon as her gland's output fell below normal. She never suffered symptoms of hypothyroidism.

Mrs. Smith now takes synthetic thyroid hormone, one pill a day. Her eyes have decreased in size considerably but are still somewhat prominent. Most importantly, she is satisfied with her appearance and feels completely well.

– END –

18. As High as a Giraffe's

The top line on Harold Boykin's emergency room ledger was cryptic: "31-yo bm w/cc severe ha and blurry vis p. wk. Hx from wife." [Thirty-one-year-old black male with a chief complaint of headache and blurry vision over the past week. History obtained from patient's wife.]

The first step in ER triage is to obtain a chief complaint and take vital signs: pulse, respiratory rate, blood pressure. Adele, the ER triage nurse, checked Mr. Boykin's pulse at a regular 80 beats per minute, respirations at 18 per minute. Both were normal. To take his blood pressure she wrapped a cloth cuff around his upper arm and placed her stethoscope bell just below the secured cuff. She placed the other end of the stethoscope in her ears and pumped up the cuff to register 200 mm Hg. [Millimeters of mercury, the units for blood pressure. Hg is the chemical symbol for mercury.]

This procedure is routine for taking blood pressure. At 200 mm Hg the brachial artery in the arm is occluded and no blood can get through. With slow deflation of the blood pressure cuff blood begins to flow through the artery and you can hear this movement with a stethoscope placed over the arm; what you hear is the gentle knocking sound of blood pushing through the partially constricted vessel with each heart beat. At *that* point the cuff pressure equals the patient's higher or *systolic* blood pressure, normally between 120 and 140 mm Hg. With further deflation of the cuff blood flows more freely through the fully-opened artery and the sounds become inaudible; *that* point represents the patient's lower or *diastolic* blood pressure, normally around 70 to 80 mm Hg.

At 200 mm Hg there should have been silence over Mr. Boykin's arm, but Adele heard the gentle 'knock-knock-knock' of blood rushing through. This meant Mr. Boykin's systolic blood pressure was *higher* than 200. How much higher?

"Let me try again," she said, and she deflated the cuff to restore the circulation. Then she pumped the cuff up to 225 mm Hg. Again she heard: 'Knock-knock-knock.'

Adele deflated and then re-inflated the cuff for a third try, this time to 250 mm Hg. 'Knock-knock-knock.' Such an incredibly high pressure! Somewhat in disbelief, Adele looked at Mr. Boykin: "Sir, are you OK?"

Tall, strongly built, with jet black hair and thin mustache, rugged,

intelligent face, Harold Boykin did not answer. He stared past the triage nurse, awake, eyes open and showing no distress but obviously unaware of her question. Mrs. Boykin, who had been standing nearby, saw the surprise look in Adele's face.

"Nurse," she asked. "What's wrong? What's wrong with my husband's blood pressure?"

Adele did not answer right away. Instead she reached for the intercom. "Dr. Randall, come to triage please. Dr. Randall, triage, *please*!"

* * *

The blood pressure of a giraffe is the highest of all animals, reaching about 300 mm Hg in its systolic, or upper phase, and 200 mm Hg in its diastolic, or lower phase.

Normal *human* blood pressure rises with age: around 120/70 for young adults, up to about 140/90 for people over 65. A blood pressure consistently above 140/90 signifies hypertension in most people. How *much* the pressure is above 140/90 is used to classify the hypertension as mild, moderate or severe, and to determine treatment.

The giraffe's blood pressure is higher than ours because the giraffe's heart has to pump blood up a neck seven feet long to reach his brain. The human brain, being only about 12 inches above the heart, is quite nicely served by a much lower blood pressure.

All blood pressure measurements are recorded relative to mercury, a dense element thirteen times as heavy as water. If your blood pressure is "120 over 70" the pressure in your arteries will support an enclosed column of *mercury* 120 mm Hg high in the systolic phase, and 70 mm Hg high in the diastolic phase. These values can also be related to water, which is much closer to blood's density than is mercury.

To summarize blood pressures in the giraffe and human:

<u>GIRAFFE</u>

- Average distance between adult brain and heart = 7 feet.
- Normal blood pressure = 300/200 mm Hg; this pressure will support a column of *water* 3900 millimeters high (12.8 feet) in the systolic phase and 2600 millimeters high (8.5 feet) in the diastolic phase.

<u>HUMAN</u>

- Average distance between adult head and heart = 12 inches.
- Normal blood pressure = 120/70 mm Hg; this pressure will support a column of *water* 1560 millimeters high (5.1 feet) in the systolic phase and 910 millimeters high (3 feet) in the diastolic phase.

Give the giraffe a human's blood pressure and the animal will die in a

state of shock, unable to pump blood to its brain. Give a man a giraffe's blood pressure and he will, if death does not come quickly, at least sustain widespread organ damage.

* * *

So-called malignant hypertension – blood pressure high enough to cause damage early in life – has a much higher prevalence in blacks than in other racial groups. The Boykin family has been particularly hard hit. Harold's father died of hypertension-related heart disease at age 58. An older brother is under treatment for the same problem. A sister was hospitalized for severe hypertension (eclampsia) during two pregnancies.

Harold became aware of hypertension at 21, his last year of Army service, when a Medic told him his pressure was high and to "take some pills." He finished his Army duty and was honorably discharged. For the next five years he attended a Veterans' Hospital medical clinic, for blood pressure checks and medication. Control of his pressure was erratic, there were frequent adjustments of medication, yet all the while he felt well. The last drug prescribed affected his libido and was his 'last straw:' Too many medications, intolerable side effects, and all for a disease that didn't make him feel bad. Disillusioned, Harold quit attending the clinic and stopped all therapy.

Twice after leaving the veterans' clinic Harold showed up in Mt. Sinai's emergency room for flu-related symptoms. On each visit his blood pressure was taken and found elevated and he was advised to attend our hypertension clinic or return to the VA. Twice he ignored the advice.

Then, at age 31, some six years after his last regular therapy for hypertension, Harold developed a severe headache. He made no connection between the headache and high blood pressure, especially since he found some relief with aspirin.

A few days later he complained to his wife of blurry vision *and* persistent headache. Still no connection. Mrs. Boykin actually called the hospital's eye clinic for an appointment, thinking her husband might need glasses. The next day his headache became intolerable and she drove him to Mt. Sinai's emergency room.

* * *

His blood pressure in the ER was finally measured at 280/180 mm Hg, one of the highest recorded in our hospital. Under Dr. Randall's direction

the ER nurses did an electrocardiogram, drew blood and began treatment with under-the-tongue nifedipine, a drug that induces an immediate lowering of blood pressure.

Fifteen minutes after receiving nifedipine Harold's blood pressure was slightly lower, 260/160 mm Hg. In another fifteen minutes it was 250/150. At that point he was transferred to MICU.

On arrival to MICU he appeared in good physical shape, albeit confused. He made no eye contact and did not talk or respond to questions, suggesting swelling of the brain, a potentially fatal condition known medically as *hypertensive encephalopathy*.

We found other stigmata of severe, sustained hypertension: tiny hemorrhages in the back of his eyes (the retina); a large, bounding heart; and protein in his urine. Neurologic exam did not reveal any evidence for stroke or brain hemorrhage (a brain CT scan was ordered to be sure). Blood drawn in the ER showed diminished kidney function and his electrocardiogram helped confirm an enlarged, hypertrophied heart muscle (cardiomegaly). The damage sustained by his eyes, heart, kidneys, and brain reflected an enormous arterial pounding: the blood pressure of a giraffe in the body of a man.

We started an infusion of *sodium nitroprusside*, the most potent anti-hypertensive agent available. Nitroprusside directly dilates the arterial blood vessels and almost never fails to lower blood pressure.

Because it is so potent nitroprusside is a tricky drug to use. If it lowers blood pressure too much the patient can go into shock from *hypo*tension. If nitroprusside doesn't lower blood pressure enough the patient will remain at high risk for stroke, heart attack, or kidney failure. The goal of nitroprusside therapy is not a normal blood pressure but one that is 'safely elevated,' something on the order of 160/100 mm Hg. The drug's manufacturer cautions:

> [Nitroprusside] should be used be used only when the necessary facilities and equipment for continuous monitoring of blood pressure are available.

"Necessary facilities" means an intensive care unit and round-the-clock nursing care. "Equipment for continuous monitoring" usually requires threading the patient's radial (or other) artery with a thin catheter. The catheter is connected via plastic tubing to an electronic monitor so arterial pressure can be continuously recorded and digitally displayed at the patient's bedside.

We began infusing two micrograms of nitroprusside per kilogram body weight each minute (notated in the chart as '2 ugm/kg/min'). Within an hour Mr. Boykin's pressure fell to 240/130 mm Hg; within another two hours, 230/122 mm Hg. Still high, but safer.

Intravenous nitroprusside is impractical for long periods and is also potentially dangerous. A metabolite of the drug, thiocyanate, is a cyanide-like compound that can starve the body of oxygen. Cases have been reported of cyanide toxicity from prolonged nitroprusside infusion. We planned only a short course.

The next day Mr. Boykin was awake but still not communicating. On a nitroprusside dose of 3.5 ugm/kgm/minute his blood pressure was down to 200/110 mm Hg, still elevated but not life-threatening. We started him on oral anti-hypertensive drugs and began lowering the nitroprusside dose.

* * *

Most of the 20 million hypertensives in the country never require hospitalization for high blood pressure, let alone intensive care. At Mt. Sinai two to three percent of our MICU patients come in for severe hypertension; these patients are usually 'new' hypertensives (previously undiagnosed and often with occult kidney disease), or 'known' hypertensives who have been non-compliant with prescribed medication.

If a patient is compliant in taking medication and receives good follow up in the clinic or office, blood pressure can usually be controlled. In fact some patients improve with just a change in life style: weight loss, no smoking, low salt intake, and exercise.

As to drugs, there are many, many choices, from the tried-and-true to the new-and-exotic. Paradoxically, an explosion in the number of medications in recent years has made treatment both easier and more complicated. Easier, because therapy can now be tailored to the individual patient; more complicated, because the sheer number of drugs makes it difficult for doctors to keep up with their nuances and side effects.

For example, captopril, a popular anti-hypertensive drug, is a member of the relatively new ACE-('angiotensin-converting-enzyme') inhibitor group of drugs. Doctors commonly prescribe captopril and other ACE inhibitors in place of an older drug type, such as a diuretic, whose side effects may be better appreciated.

ACE inhibitors have two peculiar side effects. One is a persistent cough that occurs in a small percentage of patients taking the drug; treatment of this problem is to stop the drug, but first the cough must be appreciated as a drug side effect. Sometimes patients go through many tests before the side effect is appreciated. Another, and fortunately rarer side effect of ACE inhibitors, is a sudden swelling of the face and tongue; treatment requires stopping the drug and corticosteroids and anti-histamines for a few days. If the swelling

is severe the patient will be hospitalized for observation.

Below is a list of oral anti-hypertensive drugs (by no means complete) according to drug type. Under each type the generic drug name is given, followed by brand name(s) in parentheses.

1. DIURETICS
   Furosemide (Lasix)
   Hydrochlorothiazide (Esidrix; HydroDIURIL)
   Chlorothiazide (Diuril)
   Metolazone (Zaroxolyn)

2. CALCIUM CHANNEL BLOCKERS
   Nifedipine (Procardia)
   Verapamil (Calan; Isoptin)
   Diltiazem (Cardizem)
   Nicardipine (Cardene)

3. BETA BLOCKERS
   Atenolol (Tenormin)
   Propranolol (Inderal)
   Timolol (Blocadren)
   Metoprolol (Lopressor)
   Betaxolol (Kerlone)

4. ACE INHIBITORS
   Captopril (Capoten)
   Enalapril (Vasotec)
   Lisinopril (Zestril)

5. MISCELLANEOUS:
   Prazosin (Minipress)
   Clonidine (Catapres)
   Methyldopa (Aldomet)
   Hydralazine (Apresoline)

A *single* drug is usually prescribed for mild to moderate elevations of blood pressure. *Two or more drugs* are used for severe cases or when hypertension doesn't respond to a single agent. The actual dose of any drug depends on the patient's tolerance and response. Given the type and number of available drugs, plus the range of dosages, hundreds of outpatient regimens can be formulated.

* * *

By the middle of Mr. Boykin's second day in MICU we were able to stop nitroprusside infusion and continue therapy with just two oral medications, Catapres and Lasix. His brain CT scan showed no bleeding or stroke so we expected decent recovery from the encephalopathy.

By day three his blood pressure was 170/105 and the encephalopathy had cleared. For the first time in four days Harold Boykin was alert *and* oriented. I had my first discussion with him that afternoon.

"How do you feel?" I asked.

"Much better," he said, without affect. In fact I was struck by his lack of emotion on this point, certainly none of the glad-to-be-alive attitude we see in some patients. Either he was sullen by nature or perhaps still somewhat depressed by the encephalopathy.

"Mr. Boykin, do you know what happened?"

"I guess my blood pressure was high. That's what the nurses tell me."

"How long have you known about your blood pressure problem?"

"They found it when I was in the Army. That was about ten years ago."

"Do you know how serious it is?" With that question he looked at me for a few seconds, as if to say, 'What do you think, I'm some kind of jerk?' and I felt a little self conscious asking these leading questions.

"I guess pretty serious. I wouldn't have all these tubes in me if it wasn't." (He still had an arterial line and a venous infusion catheter).

He didn't object to my questions so I decided to press ahead. "Mr. Boykin, you almost died from it. As it is, your heart is enlarged and your kidneys show damage from the high blood pressure. It also affected your brain, which is why you don't remember much about what happened."

His attitude remained rather sullen. He had evidently heard such threats before. Now I was telling him *faits accompli*; these things had happened. His other doctors hadn't been kidding all these years.

"Will they get better?" he asked, with about as little emotion as if one asked 'Where is the men's room?'

"We don't know yet. Your pressure's only been under control a short time now. We have to wait and see if any of the damage is reversible. That might take several weeks. You won't stay here in the ICU, of course. You'll have to start coming to the clinic regularly. We'll give you an appointment. By the way, why did you stop taking your medication?"

"I didn't have any to take."

"I mean several years ago, when you were being treated."

"I don't know. That was a long time ago. I remember the medicine made me sick. Sick to my stomach. I actually felt better without it."

"Your wife told me you also smoke."

"Yes, I do."

"How much?"

"About a pack a day. Maybe a little less."

"How long?"

"Since I was a teenager."

I paused for a few seconds. Staring him in the eyes, I said: "I don't know if anyone's ever told you before, Mr. Boykin, but you're a walking time bomb."

"What do you mean?"

"Well, what usually kills hypertensive patients like you is a heart attack or a stroke. You came very close to having a stroke the day you came in, do you know that?"

"Now that you tell me I do."

* * *

I don't enjoy preaching to patients. So many of them are sick because they drink or smoke too much, or use illicit drugs, or don't take their prescribed medications. My Boykin's problems, tobacco addiction and hypertension, are in theory preventable or treatable, like alcoholism and cocaine abuse. Stop drinking, stop smoking, stop abusing drugs. It all sounds so simple *in theory*. The reality is far different.

Intensive care specialists take pride in bringing a diabetic out of coma or rescuing a hypertensive from the brink of death or weaning a patient away from artificial ventilation. The pride may be justified but we should ask on each occasion: would the coma, encephalopathy or respiratory failure have occurred in the first place if there was better outpatient care, more patient education, effective drug rehab programs?

It is one thing to treat an acute, life-threatening illness and another to prevent the problem in the first place. The latter is the real challenge. Compared to providing good outpatient care for patients like Mr. Boykin, that is, compared to effective *preventive medicine*, intensive care is easy.

Followup

On the fourth day in MICU Mr. Boykin's blood pressure was down to 160/95 and we transferred him to the regular ward. He stayed another week in the hospital.

Unfortunately his kidneys were irreversibly impaired, almost to the point of requiring kidney dialysis. A hypertension specialist took over his outpatient management and prescribed a regimen to help preserve remaining kidney function. The regimen included a low-salt diet and three anti-hypertensive drugs: Catapres, Lasix, and Minipress.

Perhaps frightened by events, Mr. Boykin has become very compliant. He regularly attends the hypertension clinic and takes his medication. He

knows that blood pressure pills are the only thing keeping him from a suffering a stroke, heart attack, or life-long kidney dialysis.

-- END –

19. The Red Baron

Hemoptysis. HE-MOP-TUH-SIS. The word means "coughing up blood," one of the most frightening of medical symptoms. Five quarts of blood speed through our lung capillaries every minute, ceaselessly, until we die. The meshwork of capillaries circle and envelope each of the lungs' 300 million air sacs, so that blood is never far from fresh air. All that separates lung blood from lung air is an extraordinary membrane of microscopic thinness and gargantuan proportion. Stretched out in its entirety the membrane's surface area would cover the surface of a tennis court.

Through this diaphanous barrier gases transfer both ways. Fresh oxygen goes from the air sacs into the capillary blood, to be delivered to the rest of the body; unwanted carbon dioxide goes from the blood into the air sacs, and then exhaled. This vital transfer of gases – oxygen in, carbon dioxide out – is *the* function of our lungs.

Another, separate flow of blood delivers oxygen and nutrients to the lung tissues themselves. Thus, the lungs contain two supplies of blood, one to take up oxygen for the whole body and give off carbon dioxide, the other to deliver oxygen and nutrients to the lung tissues. This is a complicated affair, but it works beautifully. Physicians hardly ever think about the body's dual blood supply (except in medical school!). If either blood supply leaks through the capillaries the person coughs and then – there it is – bright red *blood*.

Hemoptysis is scary, but by no means is it always serious or life threatening. There are several grades. "Mild" hemoptysis may occur from just severe coughing; sometimes people "hack" so hard that capillaries rupture and spill small amounts of blood into the large air passages. The blood gets mixed with phlegm and is expectorated (doctors call this "blood streaking"). The problem invariably goes away when the coughing stops.

"Moderate" hemoptysis describes the situation when the patient coughs up a mouthful or so of blood, but not on a continuous basis. The blood may be expectorated once or twice a day. This patient will usually be

hospitalized and undergo investigation with x-rays and other tests. Frequently, a bronchoscope is inserted directly into the lungs to investigate the site of bleeding. The goal is to find out not only the cause of hemoptysis but exactly where the blood is coming from (top of the lungs? bottom?).

"Massive" hemoptysis is life-threatening. Blood is coughed up in such quantity, or so frequently, that the patient is at risk of dying from shock or from flooding the lungs with blood and suffocating. Sometimes the patient requires emergency surgery to remove the bleeding lung.

Bronchitis is the most common condition associated with mild hemoptysis. Moderate to severe hemoptysis may be due to a variety of other diseases, including blunt trauma to the chest, pneumonia, tuberculosis (TB), throat and lung cancer, pulmonary embolism and some unusual "autoimmune" diseases. In the 19th century the most common cause of hemoptysis was TB, then known as "consumption" because of the body wasting seen as the disease progressed. Since the 1950s TB has been a treatable condition and is now a rare cause of hemoptysis.

* * *

When Joseph McShane, 38, came to our emergency department (ED) one Monday in April 1990, his complaint was "I have hemoptysis and severe pain." Right away this was unusual. Patients don't use medical terms unless they are medical professionals or, as is sometimes the case, their disease is chronic and they have learned the lingo of their illness. Mr. McShane drove himself to the hospital and was in no distress despite his pain, so the hemoptysis was not considered life threatening.

Except for the most critical cases, who are whisked by ambulance attendants directly into the treatment room, ED patients are asked to state their complaint to a triage nurse. She (or he) is trained to write down the response verbatim, no editing, along with the patient's blood pressure and pulse on an intake form. The nurse is also trained to know who needs to be seen right away, and who can wait. Mr. McShane was seen right away.

(Roger Bennett, the ED physician who first evaluated Mr. McShane, later told me of his surprise at seeing "hemoptysis" on the intake form under CHIEF COMPLAINT. His first thought was that Julie Bernstein, the triage nurse that morning, must have shortened the patient's convoluted symptoms into more easily understood medical jargon.)

About 5'10", 150 pounds, Mr. McShane had the lithe build of a long distance runner. He was dressed in neatly pressed street clothes, no tie or jacket. His physique, coupled with dense straw-colored hair, blue eyes, tan complexion, slightly sunken cheek bones and a straight aquiline nose bespoke an overall healthy appearance. Looks can be deceiving, of course, and no one who recently coughed up blood can be considered in good health, at least not until the problem is resolved.

"Hello, Mr. McShane, I'm Dr. Bennett. What happened?"

"I woke up this morning and felt OK initially. Anyway, I went to the bathroom and felt some pain right here [pointing to his left lower chest], then something in my throat, like a lump. Then I coughed up a half cupful of blood. I got scared. I'm from out of town, here visiting my sister, so I don't have a local doctor. Instead I drove right to the hospital. I've had this hemoptysis before, so I knew what was happening and what to do about it." (Yes, that's exactly what he said. At that point Dr. Bennett silently exonerated his triage nurse.)

"Oh? Where? When did this happen to you before?"

Mr. McShane provided a reasonably cogent medical history. He told of two previous admissions to Mercy Hospital in his home town of Atlanta, six months apart, each lasting several days. He had undergone all sorts of tests during each hospitalization. He produced a letter typewritten on Mercy Hospital stationery, dated December 15, 1989; it was signed in longhand.

To Whom It May Concern:
Mr. Joseph McShane was in Mercy Hospital August 1989 with acute pulmonary embolism. His lung scan is abnormal and diagnostic of pulmonary embolism. For acute attacks he receives heparin and coumadin. He can have severe pain with this problem. This letter is being given to him in case he travels out of Atlanta. [Signed, Dr. Howard Lee].

[To the reader: Pulmonary embolism (PE) occurs when blood clots form in the leg veins, break off and travel to the lungs. PE can cause chest pain and hemoptysis. Heparin and coumadin are blood thinners used to prevent more clots from forming and potentially killing the patient. Heparin is given by vein in the hospital, as soon as pulmonary embolism is diagnosed. Coumadin is an oral (tablet) blood thinner, used for outpatient therapy. PE is also discussed in the story Lysis and Crisis.]

After reading Mr. McShane's letter Dr. Bennett asked: "Is Dr. Lee your regular treating physician?"

"Yes. Since the last time I was in the hospital. The first time I had

another doctor and she just diagnosed bronchitis. Gave me antibiotics, which probably did no good. I think she missed the diagnosis altogether. Then I got the same symptoms again and thank goodness for Dr. Lee, he diagnosed the problem. No doubt, he said. You have pulmonary embolism." At this pronouncement Mr. McShane winced, either from chest pain or the thought of the wrong diagnosis again being made.

"How long did you take the coumadin for?"

"In the hospital they gave me heparin, and also Demerol for the pain. After a few days the pain went away and I didn't need Demerol any more. Altogether I took coumadin for about six months, between five and seven point five milligrams a day. This dose kept my prothrombin test [a measure of proper blood thinning] in the therapeutic range."

"So when did you take your last dose of coumadin?"

"About four months ago, in early January."

"And you've had no hemoptysis since then?"

"No, I've really felt fine. Until this morning."

"What kind of work do you do?" Dr. Bennett thought Mr. McShane might be a nurse or hospital worker, given his apparent grasp of medical terminology.

"I'm a computer salesman. I was in the area for business reasons, and when I come up here I always stay with my sister and her family."

Mr. McShane's physical examination was normal except for two findings. He had a fast heartbeat, or pulse, a common abnormality seen in a hundred different conditions, including ordinary anxiety. A fast pulse is like fever; it generally means something's wrong, but the cause could be just about anything. Mr. McShane's electrocardiogram was negative except for displaying the fast heart rate.

The other abnormality was pain in his left lower rib cage whenever he tried to take a deep breath. We always ask patients to 'take a deep breath' when we listen to their lungs. He could not. Midway through a deep breath he would wince, grimace and stop. Doctors call this sign "splinting" because the chest cage is splinted by pain – the patient cannot fully expand it. The cause is usually pleuritis, which is inflammation of the lining of the lungs (the pleura).

The next test obtained in the ED was a chest x-ray. The x-ray machine is kept in a small room just off the side of the main examining area. About a minute after his x-ray was taken, and before he could put his shirt back on, Mr. McShane coughed up a hunk of bright red blood. Having no towels or sink nearby, he coughed the red glob into his shirt. Alarmed, the x-ray technician called for Dr. Bennett to come take a look. Dr. Bennett came, looked, then paged me.

I was in the ED five minutes later and found Mr. McShane holding his side, wincing, in apparent pain. I introduced myself.

"Dr. Bennett told me a little of your history. Are you in a lot of pain?"

"Yes, Dr. Martin, I am...Could I have something for the pain?"

I checked the record. He had been in the ED about 35 minutes and had not received anything for pain, which was apparently becoming more severe and frequent. I ordered an injection of Demerol.

Despite the pain and fast pulse he did not require ICU care, although if he continued coughing up blood that could change. I learned that he did not smoke or drink alcohol, and had no obvious risk factors for pulmonary embolism, such as chronic heart disease, a history of prolonged immobility, or any clotting disorder. He was married and had two children, all back home in Atlanta. He had come to the ED alone, he said, and asked that we not call any family members about his condition. He did not want to worry anyone. Such a request is not unusual, and is routinely honored unless the patient is in critical condition.

I formally admitted him to the hospital on my service. Naturally, my first thought was that he had suffered another pulmonary embolism. The sudden onset of coughing up blood and chest pain practically equals pulmonary embolism until proven otherwise. Because the treatment is not easy (long term blood thinners are tricky to manage), physicians are obligated to prove the diagnosis with some certainty; this is accomplished using one or more tests.

His chest x-ray was normal, which is not an uncommon finding in pulmonary embolism. Clots and inflammation usually cast no shadow on the chest x-ray, unlike pneumonia, which always shows up as a shadow on the x-ray film.

I next ordered some blood work, then sent him for a lung scan. A lung scan is the basic screening test by which doctors diagnose pulmonary embolism. Radioactive material is injected into an arm vein and then imaged by a scanner placed over the patient's chest; emboli in the lung show up as holes or defects in a sea of radioactive dots. (The scan is a little more complicated than suggested by this brief description; see pages 154-155.)

During the hour it took to complete the lung scan, I wondered how we were going to diagnose a new lung clot in a man who already had old clots. The scan would be abnormal, for sure, but it would be difficult to separate new clots from old. Oh, well, I reasoned, it probably didn't matter. Anyone who has a pulmonary embolism once can have it again, and coughing up blood is reason enough to resume treatment. I would have to put him back on his blood thinner, this time for an indefinite period. He would also need a letter to take back home. I had his entire hospital course mapped out, and the letter dictated in my head: 'Dear Dr. Lee,' it began. 'Your patient, Mr. Joseph McShane, was recently hospitalized at Mt. Sinai for recurrent

pulmonary emboli...'

But things don't always work out as we expect. An obvious diagnosis sometimes doesn't pan out, a safe assumption is occasionally proved wrong. Whatever the explanation for diagnostic overconfidence, suffice to say that Mr. McShane's lung scan turned out normal, pretty much ruling out pulmonary embolism. At first I didn't believe the verbal report when the radiologist called, but it was true; I looked at the lung scan myself.

I next arranged for an ultrasound study of his legs. This is a sophisticated test that can actually visualize clots in the leg veins, by sending sound waves through the skin and checking how they bounce back from veins deep within the thighs. An abnormal bounce can reveal clots in the thigh veins, and these clots are the origin of most pulmonary emboli. (Ultrasound won't pick up clots in the lungs because all the air gets in the way.)

A diagnostic ultrasound study, i.e., one showing clots in his thigh veins, would suggest the clots were about to break away; if that was the case, perhaps some clots had already traveled to his lungs and caused his chest pain and hemoptysis. This would be highly unusual given the normal lung scan, but I was fishing. True, he had no leg pain or swelling, but half the patients with leg clots have no symptoms. Finding clots in his legs would be sort of like diagnosing pulmonary emboli by proxy, especially since treatment is the same as for clots in the lungs.

But the ultrasound test was also normal. There were no clots in his legs, so we had to seek another reason for the pain and hemoptysis. The problem was that I could not think of another reason.

By now Mr. McShane had been at Mt. Sinai over eight hours and it was late afternoon. I had no more tests planned for his first day of hospitalization. Before leaving I went to see him again.

"So far your tests are normal. They suggest you don't have pulmonary emboli. That's the good news."

"And the bad news, Doctor Martin?"

"Well, there really is no bad news, except that I don't know why you are coughing up blood again. You said they diagnosed pulmonary emboli last year in Atlanta?"

"Yes, that's right. My doctor was Dr. Howard Lee, Mercy Hospital. You saw his letter. My lung scan showed pulmonary emboli, and they were fairly certain of the diagnosis. Do you think it's cancer, Dr. Martin?"

"Cancer?" I said, with some surprise. I had not even been thinking of cancer to explain his symptoms. "No, I don't think it's cancer, not at your age. First, you don't smoke. Second, your chest x-ray is clear. Nothing to suggest cancer there. However, to be certain, I suppose I really should do a bronchoscopy. Sometimes cancer can cause hemoptysis and not be visible on the chest x-ray. I'd like to do the bronchoscopy tomorrow morning. I believe you had that procedure in Atlanta, right?"

"Yes, the first time I was hospitalized, not the second time. The first time is when they said the problem was just bronchitis. It's not the most pleasant test, but if you have to do it, I'm ready. Nothing to eat or drink after midnight, right?"

"Right." This guy certainly knew the protocol.

My beeper went off about a half hour later, as I was preparing to leave the hospital. It was Mr. McShane's nurse.

"Dr. Martin, can you come up? Mr. McShane just coughed up a whole lot of blood."

"OK. Please check his blood pressure and pulse, and put him on two liters of oxygen. I'll be right up."

My patient was sitting in his bed, holding a curved plastic basin full of blood. I estimated at least 30 cc's, or about 2 ounces were in the basin. There was also a little bit of blood over his chin and lower lip. He appeared apprehensive, his face contorted in pain, and his pulse was rapid at 120/minute. A quick exam of his lungs and heart revealed nothing new.

"How do you feel? Do you have any chest pain?"

"Yes," he said quietly, with little complaint in his voice. "It came on just before the hemoptysis, all of a sudden. I'm sorry to bother you, Dr. Martin."

"Nonsense," I replied. "That's what we're here for. However, I think I should send you to the intensive care unit and do the bronchoscopy now, instead of tomorrow. I need to know the site of bleeding. You might end up requiring surgery, and for that we need to know where the blood is coming from. Does it feel like it's coming from your left lung? Can you tell?"

"I don't know...Doctor Martin. Maybe it's coming from both lungs...I just don't know. But the pain is severe, I can tell you that."

I ordered another injection of Demerol and set in motion his transfer to the ICU. I also paged one of the respiratory technicians, to help me set up the bronchoscope. With the supper time delay in transport it took us another hour before Mr. McShane was settled in MICU and the bronchoscopy could start. By then the Demerol had worked and he was calm. I also ordered injection of a sedative, to relax him for the bronchoscopy.

The bronchoscope allows physicians to look inside the lungs, much as gastroenterologists look into the stomach and intestines with an endoscope. Both techniques employ a thin fiberoptic device that is a marvel of engineering. The bronchoscope is much smaller than a gastrointestinal endoscope, only six mm wide (about 1/4 inch), two feet long and quite flexible (the formal term for the procedure is "flexible fiberoptic bronchoscopy").

We pass the bronchoscope through the mouth or nose and then into the lungs. Contained within the scope's 6 mm diameter and running the entire length of the bronchoscope are: a narrow channel for suctioning mucous or blood from the patient's lungs; fiberoptic bundles for transmitting a bright light; other fiberoptic bundles that allow us to see through the scope; and a thin wire that allows us to bend the bronchoscope tip in almost any direction. By looking through the bronchoscope we can usually tell the site of bleeding as long as it has been recent (as Mr. McShane's was).

He was cooperative during the procedure. I passed the scope with no difficulty and was able to see all the major air passages in both lungs. But I saw no blood; it was all gone. This sometimes happens. The patient coughs up what little blood leaks into the air tubes, and by the time you get around to looking with the bronchoscope the blood has disappeared, presumably all coughed out.

Well, he had surely had hemoptysis. I and the nurses saw the blood. Furthermore, he had a history of coughing up blood, and a diagnosis of pulmonary emboli was made in Atlanta. Then I began to wonder. Could the Atlanta doctors have been wrong about the diagnosis? Could they have missed another, more insidious cause, such as an uncommon autoimmune disease? Anything was possible, but given the findings to that point, I was still concerned about pulmonary embolism.

My plans were to watch him overnight and consider doing a pulmonary angiogram the next day, a test that, for all its faults, is considered the "gold standard" for diagnosing pulmonary emboli. One of the test's difficulties is that it is highly "invasive." A catheter has to be inserted into the heart's chambers so dye can be injected to outline blood vessels that serve the lungs. Clots show up nicely with this test, but the angiogram's invasiveness and expense limit its use. The lung scan is more indirect but much easier to perform. Perhaps only one out of every 100 patients initially suspected of having pulmonary embolism ends up with an angiogram.

That night, about 10 p.m., I called the ICU to see how Mr. McShane was doing. No one had called me, so I knew he wasn't in shock or deteriorating, but I did have visions of him coughing up a little more blood and of the house staff fretting over when he might open up and exsanguinate. (We had typed his blood on admission and were prepared to transfuse if necessary). One of the MICU nurses answered the phone.

"Hi Greg, this is Dr. Martin, how's Mr. McShane doing?" Mr. McShane was only one of eight MICU patients that night and not the sickest by far, but he was the most recently admitted to the unit.

There was a short pause, then a slight, sardonic laugh. "Dr. Martin, he's just fine. No more hemoptysis. His heart's beating a little fast but the rhythm's normal. The rest of his vital signs are stable." Greg didn't say it, of course, but his tone was unmistakable: 'What's this patient doing *here*? He seems to be too healthy for the intensive care unit.' But then Greg hadn't

seen him upstairs, just before the ICU transfer.

"By the way, Dr. Martin, did you know this guy was once the United States welter weight boxing champion?"

"Really?" I know next to nothing about boxing, but Greg seemed impressed, and it was a facet of my patient new to me.

"Yea," Greg continued. "He was telling us how he won the title when he was only twenty-six, in Boston, but then he gave up fighting to join the U.S. merchant marine. This guy's been around."

"So he has," I said reflexively, not realizing how ironic my comment would soon seem. "Well, let's watch him overnight. You can transfer him if you need the bed for another patient. But call me if he coughs up any more blood."

* * *

Mr. McShane was transferred out of MICU early the next morning, to make space for another, far sicker patient. By the time I reached the hospital he was already back in his regular semi-private room. After checking the MICU patients I went up to see him; it was about 8:30.

"I understand you've coughed up no more blood since the bronchoscopy. How are you feeling?"

"OK, I guess. That last injection helped quite a bit." I had left a standing Demerol order for severe pain. Since the bronchoscopy he had received one more injection.

"Dr. Martin, are you going to do the angiogram this morning?"

"I'm not sure. I want to discuss that with you. So far, everything appears normal except for your fast heart rate, which I also cannot easily explain."

"Oh, I see. But if I don't have pulmonary embolism, what is causing my hemoptysis?"

"I don't know. And I can't treat you because I don't know what to treat for. Sometimes people cough up blood and we never find a cause. Then there is a group of autoimmune diseases that are difficult to diagnose, and they can cause hemoptysis. Some of the blood samples we drew yesterday are to check for these unusual diseases, and the results should be back later today.

"The one major procedure we haven't done is a pulmonary angiogram. This involves placing a long catheter into your heart and squirting some dye through it, to directly visualize any clots sitting in your lungs. It's an invasive test and probably unnecessary in your case, since your lung scan is

normal. We usually recommend an angiogram when the lung scan is equivocal, but yours really isn't. In fact, in your situation the findings rule out pulmonary embolism with almost ninety-nine percent certainty. We'd need to do a pulmonary angiogram to make it one hundred percent, but I'm reluctant to push for an angiogram unless you understand these odds. Since it's an invasive test there are some risks, some potential complications."

I thought at the time: he *could* be one of those extremely rare individuals with a normal lung scan despite the pulmonary embolism. Faced with a strong story for embolism, and a normal scan, some doctors would just watch him without doing any more tests or giving any treatment; the assumption, entirely valid, is that if a clot is present it is just too small to do any real harm. Other doctors might opt for doing the angiogram, on the theory that it is important to make a proper diagnosis, and that maybe he should be treated even if the clot is only a small one.

It did not seem unreasonable to propose an angiogram, given the circumstances, and I was prepared to present my case. Mr. McShane was an intelligent patient with a serious problem and he should be involved in the decision making. I expected a barrage of questions and further discussion leading up to an informed consent. To my surprise, he didn't hesitate.

"I understand everything you say, Dr. Martin. Let's do the angiogram. I want to know why I'm coughing up blood."

"Are you sure?"

"Yes."

"Don't you want to know the potential complications?"

"I trust you, Dr. Martin. If you recommend it, that's good enough for me."

I wanted informed consent, not 'Yessir, anything you say, sir!' This guy was too eager. Too eager. No one has ever agreed so readily to have a pulmonary angiogram. Always, there are some questions, a quick consultation with the family, or some other hesitancy over having a catheter inserted into one's heart. I began to wonder (probably for the first time) about his psyche. He had a serious medical symptom but didn't seem to be handling it like a patient. What was the problem?

In retrospect, he was too smooth. I find this difficult to explain, but it seemed as if he was both the patient and *apart* from the patient. He had the proper symptoms, but not the proper affect. Except when he complained of chest pain, I had the unsettling feeling he could put on a white coat and play *my* role, that of the detached professional. I needed to get out of his room and think things over.

"OK," I said, "Let me see if I can set up the angiogram this morning. I'll be back to let you know soon."

I went to the nurse's station and for the next few minutes pondered the situation with his chart open in front of me. I wasn't exactly sure what to do next about making a diagnosis. There was a 99% chance that a pulmonary

angiogram would be normal. Should I subject him to this invasive procedure now, or do some other test (?what), or just wait for results of the blood tests (which, at best, would be inconclusive)? For a few minutes my mind just wandered. Then it hit me, the obvious next step.

I placed a call to the Atlanta hospital where he had been a patient the year before. Certainly a little more detail about his medical history couldn't hurt, and might even help decide what to do next. It was early and I figured the physician might be making hospital rounds.

I made the call myself and had no trouble reaching the Atlanta hospital's switchboard operator. I introduced myself and asked if Dr. Howard Lee was in the hospital, and if not could she please give me his office number. After the customary 'One moment please' there was a long pause, perhaps half a minute.

"I'm sorry, Sir," she came back, "but we have no one here by that name. You did say Dr. Howard Lee?"

"Yes. Are you sure? Has he left the hospital staff?"

"I don't know, sir. I only have a list of active staff physicians. Are you sure you have the right hospital?"

A copy of the 'To Whom It May Concern Letter' was in McShane's chart. I checked it again.

"Yes, I'm sure of that."

"Would you like me to connect you to the Office of Medical Staff? They would know if this doctor recently left the hospital."

"Yes, please."

I was connected to a secretary in Mercy's Medical Staff Office. She listened dutifully to my query, then put me on hold while she checked some records. About two minutes later she returned.

"I'm sorry to keep you waiting, Dr. Martin. We have no record of any such physician on our staff in the past five years. You say you have his letter on our hospital stationery?"

"Yes, right in front of me." I read her the heading, address and all. This bit of hard evidence intrigued her, as well it should.

"Would you mind faxing that letter to me? If you do that, I can show it to a few people and maybe help you that way."

Strangely, my first thought was that this letter pertained to a patient of mine and that I could not send any part of the medical record without his permission. But this thought quickly dissipated as I contemplated the specific circumstances and how I came to have the letter in the first place.

"Yes, I'll fax it right now. Just do me a favor, please."

"What is that?"

"I need to make some decision about this patient fairly soon. Could you work on this now and get back to me by noon? Either way, even if you come up empty handed?"

"Yes, Dr. Martin. I'll do my best to find out more about this matter. And I'll get back to you either way."

"Thanks," I said, and gave her my office and beeper numbers. It was now shortly after 9 a.m. I saw no reason to go back to Mr. McShane's room right away. I would wait until I had more information.

* * *

About 11 a.m. I was paged to the phone. It was Mercy Hospital's switchboard operator. She had a call for me from Dr. Howard Lucas, an Atlanta internist. I knew it was about McShane but my mind was a blank. What was the real story?

"Dr. Martin?"

"Yes."

"Hello. I'm Dr. Howard Lucas. Beverly [the medical affairs secretary] fax'd me your letter a few minutes ago, and asked me to call you. Hope I'm not interrupting something important." His voice was pleasantly southern, with long i's and soft consonants.

"No, Dr. Lucas, not at all. Thank you for calling. Maybe you can clear up a few things. Do you know about my patient or his letter? We admitted him to Mt. Sinai Hospital yesterday morning. He gave us that letter and said he was in your hospital for hemoptysis, and that Dr. Howard Lee took care of him. I gather there is no Howard Lee on your staff?"

Dr. Lucas let out a soft chuckle. "Yes, I'm afraid we know your patient. Beverly called the legal office when she got your fax. It seems that a patient checked out of here last year AMA [against medical advice], and before leaving he had somehow managed to lift a bunch of hospital stationery and other supplies. And his name was Shane, John Shane. Well, I was his doctor, so Beverly called me next. Is your patient about five feet ten, slim, brown hair? Sort of looks athletic?"

"Yes, that's him. Then you had him in the hospital?"

"John Shane was our patient. Came in with hemoptysis, pretty convincing story. We did all sorts of tests but found nothing wrong. Yet he kept coughing up blood. He really had us puzzled. He had a lot of chest wall pain, too. Got Demerol about every six hours until we wised up.

"About his third or fourth day here he coughed up blood while watching television in his room. He called a nurse over and without taking his eyes off the tube said, 'Here, here's some more blood' like he was giving her a urine sample or something. She thought his affect was strange, especially since she had been giving him injections for pain, and called me. By then we were mighty suspicious. I sent the "blood" for examination. Guess

what? There was no blood in the sample! It was ketchup or something. The guy's a con artist. A real sicko. We're convinced he faked his hemoptysis the entire time. Sorry you got caught with him."

"Did you call in a psychiatrist?"

"Well, we were going to, but as soon as we exposed him he signed himself out AMA. Haven't heard from or about him since. Until just now."

"Well, it certainly sounds like the same guy," I said. "He also told us he was in your hospital twice, about six months apart, and that the first time doctors just diagnosed bronchitis."

"Is that so? I'm sure we only had him once, the time I just told you about. I guess he makes up his story as he goes along."

"He also told us he's visiting his sister here, that he's a computer salesman and lives in Atlanta with his family, a wife and two kids."

"Now that you mention it, I remember he told us he was from Texas or some place out west, and was in Atlanta for an accountant's convention. I never heard about any family. I'm telling you, the guy's a looney tune. He sounds like a classic Munchausen. Didn't pay his bill, either. In fact, his insurance card was phony and all his bills were returned with No Forwarding Address stamped on them. He stiffed the doctors and the hospital."

"I think I'm getting the picture. Thanks, Dr. Lucas. You've been very helpful."

"No problem. Glad to help. Good luck with this guy. See if you can get psychiatry to see him. He certainly needs their help."

I felt like someone had punched me in the stomach, deflated my tires, stolen my bicycle. What kind of person fakes a serious illness? And why? I went back to the room to confront Mr. McShane aka Shane. Although I now felt betrayed by this imposter, I had to remind myself that he was still a patient, and would remain so until I could confirm this bizarre story and transfer his care to a psychiatrist. So, in a quiet, non-confrontational manner, I walked back to his bedside. He spoke first.

"Did you arrange for the angiogram, Dr. Martin?"

"No, I called Mercy Hospital instead. In Atlanta."

He showed no concern, no surprise. After all, he had given me the hospital's name, so I guess he figured there was nothing to hide.

"I spoke with a Dr. Lucas. There is apparently no Dr. Howard Lee on their staff."

"Oh? Well, he must have left. But it was his letter, alright."

"Dr. Lucas says he took care of you last year, but that he knows you under a different name, as Mr. Shane, not *Mc*Shane."

"Sorry doc, I changed it recently. Having some alimony problems. I

should have told you."

Lucas not Lee. Shane not McShane. And sorry *doc*? What's this *doc*? For almost 24 hours it had been Doctor Martin, formal, distancing. Now with his lies unraveling I was being addressed *doc*, and said with a touch of disdain. Oh, oh.

"He also said you claimed to be coughing up blood last year, but all they found under the microscope was ketchup, or something like ketchup."

I said these things calmly, matter-of-factly, careful not to sound like a prosecutor out to destroy the made-up alibi of some wretched defendant. My patient was sicker than anyone had realized and an accusatory posture would not help the situation. He had to know that we knew the truth and that we knew his needs were psychiatric, not medical. It would do no good to play his game, whatever that game was.

The truth was too much. At the mention of "ketchup" his head snapped into position and he stared straight at me. The intensity of his stare was frightening; if his eyes were lasers I am certain they would have burned two holes in my skull. After about ten seconds he spoke. There was anger in his voice.

"That is a bald...faced...LIE!"

The last word was yelled. I stepped back a pace, half expecting him to jump out of the bed and pummel me. (Was he really an ex-welter weight?) He started to snarl, then contorted his face to express disgust: raised upper lip, flared nostrils, gritted teeth. Would he spit next? I decided to keep silent. The ball was in his court.

He relaxed his features. Then, with contempt in his voice: "Look, doc, you're a nice guy, but if YOU can't find the cause of my hemoptysis I'll go someplace else, to somebody COMPETENT. I don't need this phony accusation stuff thrown at me!"

A passing nurse entered the room, attracted by his loud voice. I motioned her to stay behind me, that everything was OK, and then tried to calm him down. I sensed my effort would be futile.

"I'm sorry, I'm just reporting what Dr. Lucas..."

"Lucas Schmucas," he interrupted. "He's a phony, too. Cut the crap, doc. Hey, whatever happened to your HIPPOCRATIC oath? Did you lose it somewhere? Maybe I can help you find it!"

Now he was visibly agitated. He jumped out of bed on the side opposite us and fidgeted with the night stand drawers. He opened a drawer and took out his clothes: pants, shirt (blood-stained), shoes and socks, and a folder full of papers (more letters?). In another two minutes he was out of his hospital gown and dressed in street clothes.

"Where are you going?"

"Sorry doc, you guys had your chance. Hey, what kind of doctor are you, anyway?"

`"Mr. McShane, you need some help. Please stay and let us call a

psychiatrist. You can't keep going from hospital to hospital with this complaint of hemoptysis. There's nothing wrong with your lungs. Somebody could do an unnecessary procedure and you could be harmed. Let us try to help you."

"Help me? Hah! You're the one who needs help. I've NEVER seen such unprofessional behavior. NEVER. And I've seen a lot, believe me!"

I kept my distance and did not reply. The nurse asked if she should call Security. No, I said, reminding myself that we can't keep patients against their will. Security would be no use in this case.

In less than a minute our patient walked off the ward, grabbed an elevator and was gone.

* * *

Con man? Not quite. A mentally ill man is more like it. Actually, Mr. Shane/McShane is a classic example of a condition long described in medical and psychiatric circles, the Munchausen Syndrome. The syndrome is named after an 18th century German, popularly known as Baron von Munchausen, real name Karl Friedrich Hieronymus, Freiherr von Munchausen (1720-1797). Munchausen, a soldier who served with the Russians against the Turks, was notorious for telling entertaining stories of outlandish proportion, entirely made up.

Munchausen's reputation was enlarged upon by a contemporary, one Rudolph Erich Raspe, who anonymously published (in German) *Baron Munchausen's Narrative of His Marvellous Travels and Campaigns in Russia* (1785). This work, soon after translated into English as *The Adventures of Baron Munchausen*, became a popular book of the late 18th century and has gone through many editions. A copy of *The Adventures*, as well as several other books about the infamous Baron and his tall tales, may be found in some libraries.

The term "Munchausen's Syndrome" was first used in a 1951 article in the British medical journal Lancet. A group of patients repeatedly sought hospitalization by simulating symptoms of illness. Dr. Richard Asher, the author of the article, wrote:

> Here is described a common syndrome which most doctors have seen, but about which little has been written. Like the famous Baron von Munchausen, the persons affected have always traveled widely; and their stories, like those attributed to him, are both dramatic

> and untruthful. Accordingly the syndrome is respectfully dedicated to the baron, and named after him.
>
> The patient showing the syndrome is admitted to hospital with apparent acute illness supported by a plausible and dramatic history. Usually his story is largely made up of falsehoods; he is found to have attended, and deceived, an astounding number of other hospitals; and he nearly always discharges himself against advice, after quarreling violently with both doctors and nurses...

Since 1951 there have been medical reports of patients faking abdominal pain, seizures, kidney stones, back pain, asthma, mental confusion, fever, blood in the urine, hemoptysis and a variety of other illnesses. Although Asher called the syndrome common, it may only appear common because one Munchausen patient may be seen by so many different physicians and at a variety of institutions. A review published in 1967 found only 59 cases reported in the literature to that time. Munchausen remains a rare syndrome. In the 1967 review Munchausen men outnumbered women three to one; the age range was 19-62 years, with a mean age of 39.

Typical features of the Munchausen patient include:

- faking an acute illness that requires hospitalization
- familiarity with medical terms and diseases
- lack of any obvious external reason for seeking hospitalization
- aggressive behavior toward health professionals when the truth of the symptoms is challenged
- leaving the hospital against medical advice

Munchausen patients frequently travel from city to city, or even to several hospitals within one large metropolitan area. It is not unusual to uncover a string of hospital admissions within a one or two year period, and spanning several states. There is one report of a patient who had, over a 16-year period: 40 hospitalizations in three states for abdominal pain, chest pain, loss of consciousness, blood in the urine and fever – all apparently factitious; 32 emergency room visits for the same problems; four abdominal operations; and one brain operation. Whenever he presented to a hospital that had cared for him many times before he always told the doctors it was his first time there.

Another patient, a 33-year-old man, was discussed in two separate articles that appeared in a single issue of the New England Journal of Medicine (August 6, 1992). The first article covered his evaluation for a puzzling disorder at Brooklyn's University Hospital in 1991; the second article discussed evaluation for the same disorder after he was admitted to Yale New Haven Hospital in early 1992. The problem? Sudden onset of

coughing up *and* urinating blood. Doctors at Yale knew nothing about the previous evaluation in Brooklyn. At each hospital, after an extensive workup, his physicians diagnosed the Munchausen syndrome and on both occasions he left AMA after he was exposed.

Why do people fake a serious illness? No one knows for sure, but the problem is widely accepted as psychiatric in nature, a form of character disorder manifesting as antisocial behavior. Munchausen is not a psychosis like schizophrenia, and paradoxically is much harder to treat. These patients don't respond to tranquilizers or other mind-altering drugs.

Munchausen patients who have been studied by psychiatrists seem to have one thing in common: an unhappy, unloving childhood. Their early home life is often described as abusive and neglectful. As adults they become attention seeking and in this way 'act out' their rage over past deprivations. They seek nurturing for what they lacked in childhood.

Why does their nurture-seeking take place in hospitals? Because of some early experience they have learned to manipulate the medical care system. A modicum of medical training is common. Tricking doctors and nurses seems to satisfy their need to get back at society, even if this behavior puts them at risk for harm (by having unnecessary tests and procedures).

The only effective treatment, if it can be called that, is to replace their episodic (and chaotic) hospitalizations with intensive psychologic counseling in a chronic care facility, i.e., long term care in a psychiatric institution. Suffice to say, it is the rare Munchausen who ends up in such an arrangement.

William Bean, an eminent physician of internal medicine, described one Munchausen patient in verse; his poem ended thus:

I'm sorry I cannot fasten my claws on
What causes the syndrome named Munchausen
This off again, on again, gone again Finnegan
Comes in, goes out, and at length comes in again.
Munchausen's victims must be expected
To plague our lives unless deflected.
So be alert for this great nonesuchman–
Munchausen syndrome's flying Dutchman.
– William B. Bean, M.D. The Munchausen Syndrome. Perspectives in Biology and Medicine. Spring 1959.

* * *

Several puzzling questions remain about Mr. Shane/McShane. What caused his fast pulse? The heart beats involuntarily. Unless you exercise or take a stimulant, you can't accelerate the rhythm. Likely he took some stimulant to keep his heart beating fast and make us think he had a serious

medical problem. Except for this abnormality he was the picture of health, and should have had a normal or even slow heart beat (like a true athlete). We never found the stimulant, but then we didn't have a chance to look. Had he stayed around we would have ordered a blood test for various stimulants.

Why did he make up a history that was easy to verify as false? This feature is sometimes seen in Munchausen patients when the previous medical care has been in another state. Sometimes they carry medical records with them, conveniently doctored or, as in this case, wholly fabricated on stolen stationery. They probably think physicians won't check out the story in any detail.

Munchausen patients are good at faking medical symptoms and complaints. These symptoms rivet the doctor's attention, so that details about career and travels are seldom an initial concern and not something we try to verify (until our suspicions are raised, of course). I am sure his background was also fabricated, that he was not a computer salesman nor, for that matter, an ex-boxer or accountant. His slight name change from what he called himself in Atlanta may represent some private conceit; perhaps it allowed him to believe he was covering his trail, so to speak, while at the same time keep the story straight in his own mind.

Where did the blood come from? We never knew, but then we never analyzed the stuff he coughed up. Hemoptysis is relatively common and Munchausen Syndrome is very rare, so when someone coughs up red mucous we don't ask, 'Is he faking it? Is this really blood?' Most likely Mr. McShane kept a supply of fake blood in his clothes and produced it at will. Probably not ketchup, per se, but the same stuff used in the movies to simulate bleeding.

Finally, where is he now? In typical Munchausen fashion, he has likely gone to other hospitals and fooled other doctors. As expected, the number he gave for his "sister" drew a "not-in-service" response. The insurance information he provided the hospital was phony and his medical bills went unpaid.

-- END –

20. "Mommy, why don't you hug me?"

Thirty-five-year-old Naomi Benedict was sitting in a chair at home, recovering from the flu, when she felt a sudden tingling in both legs. She stood up to stretch, lost her balance and promptly fell to the floor.

The maid ran in from the kitchen. What was wrong? Mrs. Benedict said the only problem was her legs, they felt weak and tingly as if their circulation was cut off. Otherwise she felt fine. The maid helped her stand up, then climb the stairs to the bedroom and get into bed.

It was two in the afternoon on Tuesday, March 7. Mr. Benedict, an attorney, was at work and their two young children were in school. At three o'clock she was to attend the city's Arts Council meeting, her first time out in a week. Instead of getting dressed she lay in bed, unsure what to do.

She called her husband. Julian Benedict thought her leg weakness was probably from the flu and sitting too long in one position. He suggested she call Dr. Cooper and not try to make the meeting.

Dr. Cooper, one of the town's leading internists, knew the Benedicts well as both their physician and friend. He advised her to stay in bed; if not substantially better in the morning, after a good night's sleep, she should come to his office.

The next morning I was in MICU making rounds, when Dr. Cooper phoned.

"Larry, I've got Naomi Benedict in my office. You know who she is, don't you?"

"Sure," I said, "her husband is Julian Benedict, right? I've seen their picture in the magazines." I did not know them personally but I knew *of* them. At the time he was a rising young lawyer, famous after his defense of a business tycoon charged with murder. When his client won acquittal Julian Benedict's name was all over the papers. At age 39 he was a legal star.

I was aware of Naomi as a young socialite and heiress to a family fortune made in the steel industry. Like many young and wealthy women she was active in prominent charities, one of which made donations to Mt. Sinai Hospital. She and her husband were also well known, at least locally, because of their home. 'La Maison Magnifique', so dubbed by an overly enthusiastic editor, was featured in the city magazine four months earlier. The Benedicts had spent a fortune redecorating an old French chateau-style mansion into something that, even by European standards, was stunning (at least from the pictures; I have never been inside). Naomi's college degree

was in art history and she had orchestrated the entire project.

"Well, I've been treating her for a viral gastroenteritis the past week," Dr. Cooper continued. "She had the flu with some diarrhea and was getting better, at least until yesterday. She had a little leg weakness yesterday and this morning can hardly walk. I'm not sure, but it might be progressive. She also has some diminished breath sounds. I did a vital capacity [a measure of lung function] in the office and it's down to about 70% of predicted. Larry, I'm worried about her. I'd like to put her in MICU if you have a bed."

Dr. Cooper is known for good medical judgment so if he worries about a patient I do too. "Of course," I said. "We'll get a bed ready for her. When do you think she'll be here?"

"Well, Julian's with her and he'll bring her down to the hospital. With her difficulty walking, it'll take them about half an hour."

"OK. We'll be ready."

A few minutes later Harold McAllister, Mt. Sinai's Director of Neurology, stopped by. Just past 40, tall and aristocratic in bearing, Harold is a neurologist's neurologist, which is to say he regularly consults on the rich and famous. Even in the era of CAT and magnetic resonance scanners, neurologic diagnosis is an art based on thorough history and detailed physical examination. Dr. McAllister is an artist.

"Hello Harold, what's up? Who are you here to see?"

"No one just yet. I understand Naomi Benedict is being admitted soon."

"Yes, that's right. Did Dr. Cooper call you also?"

"He left a message with my secretary to see her right away. When do you expect her?"

"Within the hour. I just talked to Cooper a few minutes ago."

"OK. Would you please page me as soon as she arrives?"

"Sure."

A few minutes later the Chief of the Department of Medicine walked in. He pulled me aside from rounds, to speak privately.

"Larry, Naomi Benedict is being admitted to MICU. She apparently has. . ."

"I know," I interrupted, somewhat surprised by this unexpected visit from my boss. "I spoke with Dr. Cooper a while ago and Harold McAllister was just here also."

"Well, I just came to tell you about Julian, her husband. I don't think you know him. He's on the hospital's board of trustees. (Dr. Cooper hadn't mentioned this.) He's one of the nicest guys you'll ever meet, a tiger in court but outside the courtroom he's a real gentleman. Naomi works on the hospital's auxiliary. I can't tell you how much money she's helped raise for Mt. Sinai. I know them both. Don't be intimidated by their wealth or position. They are down-to-earth people. She, especially, is a doll. I sure hope she's OK. Maybe Cooper is just being over cautious. Anyway, call me if you need assistance of any kind. We want to do everything necessary to

help her."

I wanted to reply that "everything necessary" is what we do for all our patients, but instead just thanked the Chief for his well-intentioned advice. Some degree of anxiety is natural when VIPs (or their relatives) become patients. Socially prominent people, including politicians, business leaders, and movie stars, always expect (and usually receive) special treatment. The difference is in nuances of service, not in basic medical care. In fact, doctors sometimes have to be careful not to let 'VIP care' affect sound medical practice. A desire not to intrude, not to bother, can sometimes inhibit doctors from doing an important test or procedure, although this is less likely to happen in MICU. I learned long ago that all patients, whether dope addict or corporation president, or the rich wife of a hot shot trial lawyer, want (and deserve) the same thing: good medical care delivered in a friendly and compassionate manner.

A few minutes later the swinging doors to MICU opened and an attendant ushered in Mrs. Benedict in a wheel chair. Julian Benedict walked beside her. From TV and magazine photos I recognized them right away, although Julian appeared shorter than I had imagined. A three piece business suit covered his stocky, muscular frame. Clean shaven, tie perfectly knotted, he appeared ready to go to court. Naomi, also smartly dressed in street clothes, carried a purse on her lap.

Unlike most patients admitted to MICU she was fully alert and in no distress. My first thought was that perhaps she didn't need to be in MICU, that she might be more comfortable in a private room in one of the hospital towers. My second thought was that the towers are not equipped to closely monitor patients, and if Dr. Cooper wanted close observation she probably should be in MICU.

The MICU nurses went right to work. Since the Benedicts had bypassed normal admission procedures they sent Mr. Benedict to the admitting office, to provide insurance information and sign some papers. Then they took Mrs. Benedict into room 2 and exchanged her street clothes for a hospital gown. After a few minutes Emily, one of the RN's, came out to get a bed scale. She gave me a knowing smile.

"Why the look?" I asked.

"Anne Klein skirt and blouse? Gucci shoes and purse? Can you believe it?"

I acted dumb. "Is that fancy?"

A rhetorical question, at least for Emily. She changed her expression to show that I was quite out of touch, and returned to Mrs. Benedict's room.

Since Mrs. Benedict could not stand up the nurses measured her height lying supine (five feet five inches). Next, they recorded temperature (normal), blood pressure (125/72), heart rate (105 beats per minute), respiratory rate (18 breaths a minute – normal), and skin turgor (normal); hoisted her on a stretcher over the bed to obtain her weight (129 pounds);

and attached EKG monitor leads to her chest. From that moment her heart rate and rhythm were continuously displayed on a bedside monitor.

When the nurses finished I put in a page for Dr. McAllister and, accompanied by Janice Dover, one of the MICU interns, went in to see our new patient. Even without the fancy clothes she was so different from our usual intensive care patient (elderly or debilitated or acutely ill). Here in MICU was a very attractive woman: light complexion, brown hair combed straight back, little makeup, with the poise and bearing of a top fashion model. Even in the bare cloth of a hospital gown, without jewelry, she looked elegant, not unlike Jackie Onassis at a similar age.

"Mrs. Benedict, I'm Dr. Martin and this is Dr. Dover. I run the ICU and Dr. Dover is one of our interns. Dr. Cooper called me when you were in his office and told us something of your problem." She smiled, then spoke with a mixture of embarrassment and concern. "Hello. I feel so foolish being here. Do you really think the intensive care unit is necessary? I don't feel sick."

"Well, Dr. Cooper is worried about your sudden weakness."

"I know. I just can't walk. I feel so helpless. What do you think it is?"

"We don't know yet. He wants us to watch you for a day or so and run some tests. If there is no progression of the weakness you'll go home or to another part of the hospital to recuperate. He's also asked the head of our Neurology division to see you. That's Dr. McAllister. He will be down in a few minutes."

"Can my husband come in for just a minute? I want to tell him not to wait around. It's not necessary."

I checked her vital signs from the nurse's records. They were all normal except for the slightly fast heart rate. "Sure, I'll go see if he's through in the admitting office."

Julian returned to MICU and went in to see his wife. He came out five minutes later and announced he was leaving the hospital for a few hours. Apparently she had insisted that he go to work and come back in the afternoon, when more would be known about her condition. There would be no problem with the kids, since the Benedicts had a full-time housekeeper.

About this time Dr. McAllister returned to MICU. Consultants on new admissions don't usually appear until after the intern has examined the patient, but in this case Dr. McAllister was asked to get involved right away. He apologized for "intruding" so soon, and suggested we interview her together, then let him do the neurologic exam. This approach was reasonable since it would obviate repetition of her medical history.

Dr. McAllister introduced himself and began eliciting her story, the outline of which is related above. Mrs. Benedict had no trouble speaking or recalling events leading up to hospitalization. She impressed us as bright and articulate and, for all the press hype, remarkably free of affectation, an ordinary (if rich) human being who wanted nothing more than to get better

and go home. On a personal level I felt sorry for her.

Accustomed to all the druggies, alcoholics, and non-compliant patients we routinely see, as well as those in coma or serious distress, her presence in MICU seemed anomalous. She was not even sick. I couldn't get the idea out of my head: what is she *doing* here? Her weakness could progress, but in the scheme of things why should it? She hadn't done anything 'bad' to justify a serious illness. No drugs, no alcohol, no promiscuity.

But of course these were foolish thoughts. There are many more diseases than those brought on by self-abuse. I reflected on Eric Siegel's *Love Story* and its depressing ending: innocence and happiness dashed by cruel fate. Mrs. Benedict was innocent. She had caught a virus and developed muscle weakness. Now, she could crash just like any other patient with a bad disease.

Another question from Dr. McAllister interrupted my ruminations. "Mrs. Benedict, did you run a fever when you had the flu?"

"I took my temperature only twice. Once, at the beginning of the flu, it was a hundred and one. Two or three days later it was down to one hundred. I haven't taken it since but I think I'm recovered from the flu. At least I feel better."

"Before you developed leg weakness, that is before yesterday, did you have any numbness or tingling of your arms or legs?"

"No. The weakness and tingly feeling came on at the same time."

"Have the numbness and tingling persisted?"

"Yes. My legs have a tingling feeling now, like they're still asleep."

"But you have sensation in your legs? You can feel the bed sheets?"

"Yes, but my legs feel almost like they're not part of me, like they're still asleep."

"Have you had any trouble breathing?"

"No. Well, wait a minute. Yesterday I did feel a little short of breath after climbing the stairs. And this morning, getting into the car, I guess I was a little winded, but that's all. It's probably because I feel so tired."

"Do you smoke?"

"No, not at all"

"How about your husband?"

"Smoke? No, Julian doesn't smoke either."

"Have you ever had any respiratory problems, pneumonia, asthma, any lung trouble before?"

"No, nothing. This is actually the first time I've been in the hospital since Kevin [their younger child] was born. That was six years ago. In fact, I felt great until last week."

"Do you do any regular exercise?"

"Yes! Doubles tennis. Twice a week."

"When was the last time you played?"

"Two weeks ago. I haven't played since I got the flu."

"There was no fall-off in your game? I mean, the last time you played, did everything seem normal in your game?"

Here she gave a little laugh. "Yes. I mean, we lost, but I felt fine."

"Other than the weakness in your legs, have you noticed weakness anywhere else?"

She opened and closed her fists a few times. "No, I feel fine everywhere else."

"Has your weakness increased since yesterday? Is it harder for you to walk this morning than when you first noticed the weakness?"

"Yes. Yesterday, even after I fell on the floor I could walk a little. This morning my legs were weaker. Julian had to practically carry me into and out of the car. I could not make it by myself. Now I couldn't walk if I had to."

"Have you had any problem with your period? Is it regular?"

"Yes."

"Any history of kidney or heart disease?"

"No, none."

"How about arthritis?"

"No."

"Do you take any medication?"

"No, only what Dr. Cooper prescribed last week."

"What was that?"

"Erythromycin [an antibiotic] and some Donnatal [an anti-spasmodic for the diarrhea]."

"You've taken no other drugs the past few months?"

"Oh, an occasional aspirin. But nothing else."

"Have you eaten anything rotten or that tasted rotten in the past few weeks?"

"No. Nothing. Do you think this could be food poisoning?"

"Probably not. Botulism – and I *don't* think you have botulism – can sometimes present with progressive weakness."

There were several more questions about possible exposure to insect spray, toxic chemicals, and other people with similar afflictions, but the answers were all negative. In essence her history was straightforward: a healthy and active woman; flu-like illness for about a week; development of abrupt leg weakness; progression over the ensuing 18 hours.

Next came the neurologic exam. Dr. McAllister was a marvel to watch as he tested her sense of smell and taste, eye movements in all directions, strength of muscles from neck to toes, reflexes, and numerous other aspects of nerve function. His exam was meticulous, thorough, and kind. He explained any poke or prod that might cause some discomfort. (He did not concentrate on her heart, lungs, or abdomen, as these organ systems are not part of a neurologic exam; Dr. Dover and myself would return to check these areas.)

Apart from her lower extremities the neurologic exam was "unremarkable." Her legs revealed the problem. Normally we can push our toes down (a motion called 'dorsiflexion') with great strength, as when pushing against a bed board or standing on our tip toes; Mrs. Benedict could not push down at all. While lying flat we can lift our legs into the air and hold them up for at least a few seconds. She could not lift her legs even a fraction of an inch. We can easily take one leg and cross it over the other, from knee to ankle. This, too, she could not do. All she could accomplish with either leg was a slight rolling motion on the bed sheet.

She had sensation in both legs and could feel Dr. McAllister's warm hands and the gentle prick of his safety pin. Also of diagnostic importance was the *absence* of muscle reflexes in her legs. Normally there is a reflex jerking of the leg if the knee is tapped with a rubber hammer. Her legs didn't budge.

After the history and exam, which took about an hour, we thanked her and went to the nurses' station.

"What do you think?" Dr. McAllister asked Dr. Dover.

"Well, she can't dorsiflex [push down] her feet, so there's weakness there. It looks like a primary motor weakness of the lower extremities."

"Exactly," he commented. "And coming on a week after a viral illness, it makes you think of one particular diagnosis."

"Guillain-Barré syndrome?"

"Very good, Dr. Dover! Yes, she has the classic picture of Guillain Barré syndrome. The weakness began in her feet and seems to be progressing upwards. Now her thigh muscles are weak. I'm also a little worried about her breathing. Larry, I didn't detect any respiratory muscle weakness but that can happen if this progresses any further. We should check her vital capacity several times a day. Until we see which way she's going I definitely want her to stay here. In the meantime we'll need to do an LP [lumbar puncture] and an EMG [electromyogram] to help secure the diagnosis."

* * *

Georges Guillain and Jean A. Barré, two early 20th century French neurologists, were among the first to describe inflammation of the peripheral nerves leading to paralysis. They recognized that the paralysis occurred most commonly after a respiratory infection. Today the eponym 'Guillain-Barré' is widely used for the syndrome of post-infection paralysis.

In GBS myelin sheaths covering the motor nerves are damaged. An analogy is the rubber sheath around a thin piece of metal wire that, when damaged, prevents the wire from transmitting electricity. In GBS the myelin sheath is damaged and the nerve does not transmit impulses. The specific mechanism is unknown, although it is probably related to antibodies

generated by the infectious agent (usually a virus).

GBS afflicts men more than women, and can strike at any age. The most benign cases show only minor muscle weakness and then remit altogether. The most severe cases go on to total paralysis. If the myelin sheaths regenerate (which happens most of the time), and the patient doesn't die from acute respiratory or cardiac failure, the prognosis for recovery is good. Joseph Heller, the celebrated author of *Catch 22*, developed severe paralysis from GBS and recovered.

GBS classically presents as *ascending paralysis*, meaning it starts in the legs and progresses up the body. Atypically, paralysis can start in the head (with facial and eye muscle weakness, for example) and *descend*, or start in the middle of the body (arm weakness) and travel both ways. The worst fear is paralysis of the respiratory (breathing) muscles. To have some idea what this type of respiratory failure is like do the following. Stop breathing without 'holding your breath.' Instead, keep your mouth and throat open but do not move your chest (rib) cage. Keep your chest perfectly still. When you can no longer do so, notice how much your chest cage moves as you take in the very next breath. If you couldn't move your chest you would asphyxiate (in about four minutes).

A totally paralyzed patient cannot breathe because his chest cage doesn't move. Without movement there is no expansion of the lungs and without expansion no fresh air can enter the blood. All totally paralyzed patients require artificial ventilation, for as long as the paralysis lasts.

Examination of the spinal canal fluid – the clear liquid that bathes the spinal cord – can help secure the diagnosis of GBS. Spinal fluid is removed for analysis through a hollow needle inserted in the middle of the back at the level of the hips. (Reach around your back to the spine just between your hips. With the tips of your fingers feel the protuberances of the spinal column. In a spinal tap the needle goes between these two protuberances.) A few drops of spinal fluid are removed and sent to the lab for analysis of protein, glucose, and cell count. A spinal tap is technically not difficult and is often done by house staff.

* * *

"I better do the spinal tap," said Dr. McAllister, "but it doesn't have to be done right away. Why don't you finish your workup, then call me. By the way, I met Mr. Benedict on his way out. I'll talk to him again when he comes back this afternoon."

Dr. Dover and I returned to finish the physical exam. Apart from the neurologic system everything was normal. We also drew blood for routine tests and did the vital capacity measurement. Our exam and tests took about 45 minutes.

Afterwards I paged Dr. McAllister. He returned to MICU and obtained

permission from Mrs. Benedict for the spinal tap. The major complication, he explained, is 'post-spinal' headache, which occurs in perhaps 15% of patients.

While she lay on one side he injected a local anesthetic into a small area of skin over the spinal column. In a few minutes the area was fully numb. He then inserted an .18 gauge spinal needle into the space between two vertebral bodies. The needle entered her spinal canal with a slight 'give.' Success. Out flowed crystal clear spinal fluid. One...two... three...four cc's. The precious fluid was collected and the needle withdrawn. Mrs. Benedict reported no pain from the procedure.

"I'll check the lab results," he said, "and call Dr. Cooper to let him know how she's doing. In the meantime, please page me when Mr. Benedict arrives. By the way, were you able to get the vital capacity?"

"Yes. It's slightly low," I said. "Three point two liters. We also did an arterial blood gas, which is normal."

In four hours we had accomplished a battery of tests and exams that would have taken much longer on the general medical ward. We had also made a tentative diagnosis and plan of action, and set up a chart of items to follow her course: muscle strength of upper and lower extremities; vital capacity; respiratory rate; body temperature; blood pressure; and heart rate. Those first few hours convinced me she was in the right place. Ascending paralysis can move fast. Doctors and nurses have to be prepared to move faster.

* * *

She went rapidly downhill. By four p.m., slightly over 24 hours after the onset of leg weakness and six hours after arriving in MICU, she began losing strength in her arms. More ominous, her respiratory rate increased to 28 breaths per minute and vital capacity fell to 2.1 liters. I met with Dr. McAllister.

"What do you think?" I asked.

"She's progressing, no doubt about it," he said. "I'm going to start plasmapheresis right away."

"Can that reverse such a rapid slide?"

"Sometimes yes, sometimes no. It's best if plasmapheresis is started before the patient ends up on a ventilator. The sooner the better. There was a large-scale study a few years ago on plasmapheresis in GBS. Something like 250 patients [Neurology, August 1985, volume 35, pages 1096-1104]. They found definite improvement in patients who received plasmapheresis. But you have to start it early in the course."

"Aren't you surprised by how fast she's progressing?"

"Yes I am. GBS usually progresses over a few days or weeks, rarely over a few hours. I've seen one other patient progress this fast. And there

are some cases like this in the literature. Cooper sure knew what he was doing by putting her in MICU."

Dr. McAllister called the plasmapheresis service. The physician in charge agreed with the plan and arranged for technicians to begin pheresis that same evening.

They didn't get a chance. As Dr. McAllister was hanging up the phone an alarm went off in Mrs. Benedict's room. We ran in. Her pulse was 160 and she was *very short of breath*.

"Dr. Martin!" she gasped.

I was shocked by the change. Neck muscles contracted with each breath and her skin was mottled and blue. Her speech was short, interrupted, gasping.

"What's.....the matter?.....What's happening to me?.....Why can't I.....breathe?"

Paralysis had ascended so rapidly that her diaphragm, the major breathing muscle that sits between the abdomen and chest, was now totally paralyzed. She was breathing only with 'backup' neck muscles, and those were about to fail.

I spoke quickly to the nurse. "Please hand me the Ambu bag. And call anesthesia. She needs to be intubated right away!" I began manual Ambu ventilation with 100% oxygen through a tight-fitting face mask. This would keep her going until we could get her intubated.

"Just breathe through this mask, Mrs. Benedict. You'll be fine," I reassured her.

Patients who can't breathe because of *lung* disease usually flail their arms and move their chest cage rapidly in and out. Mrs. Benedict did not have lung failure; her lungs, the organs of respiration inside the chest cage, were normal. Her respiratory muscles had failed and she could not expand her chest. There was *no* movement of her chest (except when some air was pushed in by my squeezing the Ambu bag). Without artificial breathing assistance she would die.

The anesthesiologist came right away and intubated her with a foot-long, 1/4-inch-wide plastic tube; one end of the tube stuck out from her mouth and the other end disappeared inside her throat. With the tube in place we had a secure airway and as soon as we connected the tube to the ventilator she 'pinked up.' The ventilator – replete with alarms and constantly monitored by technicians – was now her life support.

A quick physical exam uncovered no permanent damage. Blood pressure, heart rate and skin perfusion were all reasonable. But what a close call! My patient had almost died and, what's more, I felt certain *she* knew it.

I checked an arterial blood gas, which was adequate, then called Dr. Cooper and Mr. Benedict. Both men said the same thing: "I'll be right down."

* * *

Julian went in to visit his wife. I also went in the room, mainly to observe the cardiac monitor in case there was an autonomic surge. Despite modest sedation Naomi's mind was completely intact. The distress of being fully aware yet unable to speak or move could lead to tachycardia.

"Naomi, this is Julian. Can you hear me?"

For the moment she lay there, motionless, eyes closed, a beautiful woman with a tube in her throat, surrounded by machines and monitors and wires.

"Julian, give her a gentle nudge," I said.

He touched her shoulder. "Naomi, this is Julian. Can you hear me?"

She opened her eyes and nodded her head, slowly.

"Hi, honey. The kids send their love. I told them you're doing fine. It's five o'clock now. They're home with Gertrude, eating supper."

A tear came to her eyes.

"We miss you, honey. You'll be home soon. The doctors say this paralysis is a short term thing, that it's completely reversible. Do you understand what I'm saying?"

More tears. And in Julian's eyes, too. I wiped Naomi's tears away with a towel. It was an awkward moment and I wanted to leave them alone. Her heart rhythm looked stable on the monitor so I left the room. Julian emerged about ten minutes later, his face freshly washed. Neither of us said a word.

Just then Dr. McAllister appeared. The three of us went into a MICU conference room, a small, square space almost completely filled with a round wood table. I hastily moved books and journals off the table. We sat down and I led off the discussion.

"Obviously her condition has progressed, much faster than anyone expected. Unfortunately one of the worst things has happened, the paralysis has affected her breathing muscles. She is stable but right now can't breathe without the ventilator. We plan to start a treatment called plasmapheresis, to wash out antibodies from her blood."

"What's that?" he said quickly.

Dr. McAllister answered. "As Dr. Martin said, it's a technique that removes plasma and gets rid of antibodies that might be damaging her nerves. We think antibodies against the nerves are responsible for destroying the myelin sheaths or nerve coverings. We wash out the antibodies with a series of plasma exchanges over two weeks, five or six treatments total. It's fairly safe and there are relatively few complications."

"Does she have to receive blood transfusions with this?"

"No, not at all. We infuse albumin to replace the plasma that's removed. There's no risk of AIDS or hepatitis, if that's what you're concerned about."

"And you say two weeks?"

"Yes. That's the standard length of time most patients are treated. We probably won't see much improvement before two weeks either."

Mr. Benedict did not react to this information. Instead he looked at me and asked, "Dr. Martin, you said 'one of the worst' things. What's the worst?"

"Well, people can die from Guillain Barré syndrome. I think you know that. It usually happens not from the paralysis itself, because we can support her breathing indefinitely, but from what we call autonomic dysfunction. The autonomic part of the nervous system controls things like heart rate and blood pressure. We don't understand why, but sometimes people with this condition can have a sudden surge of adrenalin. This can cause severe blood pressure swings and cardiac arrhythmia. If there is no autonomic crisis, and her nerve coverings regenerate as they usually do, she can fully recover. That's what we're aiming for, of course."

Mr. Benedict addressed Dr. McAllister. "I know her breathing is impaired but is there any sign of recovery in the rest of her body?"

"No, but that's not surprising. It's usually progressive paralysis and then recovery, rather than some areas getting better while others get worse. Right now I'd say she's nowhere near the recovery phase. We probably won't see any definite improvement for a week or more, even with plasmapheresis."

"I see," he said. "Well, the kids want to see their Mom. I told them they can't see her now because she has a bad infection and they might catch it. I really don't want them to see her like this, but if she improves can they visit her here?"

"Sure," I said, "but I agree now's not the time. It would be much better when she's more awake and can interact with them. In fact, if there is no dramatic improvement in the next two days I'm going to recommend a tracheostomy. That will allow us to take the endotracheal tube out of her mouth and put it through a small hole in her neck. Then she can eat, smile, and stay on the ventilator as long as necessary."

"She might not be able to eat just yet," interjected Dr. McAllister. "Sometimes this condition can affect the swallowing muscles. I wouldn't be surprised if hers are already involved, the way this thing has progressed."

He was right, of course. I should have thought of swallowing difficulty before saying she could eat with the tracheostomy. "That's true," I corrected. "But I would still recommend a tracheostomy for reasons of comfort."

"Well," replied Mr. Benedict, "let's cross that bridge when we come to it."

Plasmapheresis was started that evening. There was no immediate response (none was expected so soon) and she remained ventilator-dependent. In fact, her paralysis progressed and by the next morning she could not move even her head. Only her eyes moved. We took advantage of

this last vestige of motor function to teach her to answer 'yes' (eyes up and down) and 'no' (side to side). Dr. Dover inserted a thin stomach tube through her nose to be used for feedings. Other tubes already inserted included large intravenous lines for the plasmapheresis and a bladder catheter for collecting urine.

There was no change in her condition after 48 hours. I called Mr. Benedict about the need for tracheostomy and he gave permission. The operation was done on Thursday afternoon by a surgeon. She returned from the operating room with the tracheostomy tube in place and her face free of any encumbrance.

By the end of the third day we had results of several tests, all of which were consistent with GBS. The spinal fluid protein was slightly elevated. Since the protein content reaches a peak value several days into the illness, Dr. McAllister thought the small rise merely reflected an early measurement. An EEG or electroencephalogram, done the day after she "crashed," was normal except for some mild sedative effect, also consistent with GBS. The EEG tests brain wave activity and not peripheral nerves impulses, as does the EMG. Her EMG showed normal muscles but abnormal nerve conduction within the muscles, an impairment typically seen in GBS.

* * *

On the evening of the third day, about one hour after her second plasmapheresis, the nurses turned her to change the bed sheets. Almost immediately Mrs. Benedict's heart rate increased from 95 to 160 per minute and blood pressure bottomed out at 65/30. The nurses quickly rolled her back and pushed a button to lower the head of the bed to the Trendelenburg position. (Named after the German surgeon Friedrick Trendelenburg. In this position the body is angled about 30 degrees with the head down, to facilitate blood flow to the brain.)

The MICU resident reacted swiftly to the autonomic crisis. She ordered a "wide open" saline infusion and intravenous verapamil, a drug that can slow a too-rapidly beating heart. These measures worked and in five minutes her blood pressure was up to 110/64 and heart rate down to 112 a minute. I was not in MICU at the time but it didn't matter; I could not have done a better job. (For the next few days the nurses were advised to turn Mrs. Benedict very slowly and with an eye on the monitor).

On Saturday, March 11, Dr. McAllister and I again met with Mr. Benedict. "As you can see she's doing about the same," I said. "Since last night's episode of low blood pressure she's been quite stable. She still needs the ventilator and will probably stay on it for some time."

"We're going to continue the plasmapheresis another week to ten days," added Dr. McAllister. "By then we should see some improvement, although I must tell you I have seen paralysis continue for months before there is

significant nerve sheath regeneration."

Julian listened intently, then spoke. This time it was he who had something to tell us. "A member of my firm did some work for one of the New York hospitals. He made some inquiries when I told him about Naomi's condition and obtained the name of a neurologist there. I hope you two don't mind if this doctor consults on Naomi."

"Not at all, not at all," said Dr. McAllister, mildly surprised.

"It's certainly OK with me," I added.

"Who is it?" asked Dr. McAllister.

"A fellow named X_____."

At this Dr. McAllister raised his eyebrows. "Well, you certainly picked a winner. He's probably the world's top authority on GBS. I've heard him speak several times."

"Good. I didn't know you knew him, but I'm glad you have no objection. I trust you guys implicitly. I wouldn't be on the Board if I didn't think this is a damn good hospital. I have to do this for myself. If for some reason Naomi doesn't make it I want to know I did everything possible."

"I understand perfectly," said Dr. McAllister. "When's he coming in?"

"Tomorrow. He could only come on Sunday. I'm picking him up at the airport tomorrow morning."

* * *

That afternoon I went to the library and read most of Dr. X's recent papers on Guillain Barré syndrome. Although our specialties were different I didn't want to seem ignorant of his work. His publications were mostly clinical, dealing with natural history of the disease, effect of various treatments, and long term follow up of patients. In these areas he was an authority on GBS.

There was no change in Mrs. Benedict's condition all day Saturday. On Sunday Mr. Benedict and his New York consultant showed up in MICU about 11 a.m. Dr. X, about 50, was wearing a casual sport coat without tie, and comfortable loafers. His personality seem to match: low key, self-assured, friendly. The only thing he brought with him was a large brief case full of neurologic testing equipment. I wondered what he was charging for this out-of-town visit, on what was most likely his day off. Whatever the fee, it probably didn't matter to Mr. Benedict.

I handed him Mrs. Benedict's hospital chart and x-ray folder. "I'll be in the hospital," I said. "If you have any questions just ask one of the nurses to page me." He thanked me graciously and went to read her file.

Dr. X spent much of Sunday afternoon in MICU, examining Mrs. Benedict and reviewing the hospital record, then talking with me, Dr. McAllister and Mr. Benedict. In essence he agreed with our evaluation and plans. As far as he was concerned she had a confirmed case of Guillain

Barré syndrome.

The consultant recognized his main job was to reassure Mr. Benedict about our diagnosis and medical management. He was not hired to educate him (or us) about GBS. But the lawyer in Mr. Benedict wanted his money's worth. He questioned his consultant like a star witness. It was all very cordial, even if the discussion at times sounded like a legal deposition. Dr. X, for his part, gave the information asked for.

Mr. B. "Of people who get this Guillain Barré, how many die from it?"

Dr. X. "Overall mortality is about 3%. That's in our experience and in other large series as well."

Mr. B. "What do the patients die of?"

Dr. X. "Three things, mainly. Heart disease, for one. This is usually associated with autonomic dysfunction, such as arrhythmia. Another cause of death is pulmonary embolus, which is when a blood clot breaks off from the legs and travels to the lungs. The clot comes from lying in bed so long. To some extent this can be prevented by giving small doses of heparin. I should add that her doctors have given this treatment all along." (At the mention of "her doctors" Mr. Benedict gave a slow and approving nod.) "The third major cause of death in our patients is infection, such as pneumonia or septicemia. So far there seems to be no evidence for any of these problems in Mrs. Benedict."

Mr. B. "Is there any way to prevent the other two complications, the heart disease and infection?"

Dr. X. "Only by good care and catching the problems when they arise. One area of our research is the autonomic heart problem. So far we haven't found a way to predict who will develop it or why. That's why it's so important to watch GBS patients closely, so you can treat the blood pressure crisis or arrhythmia as soon as they occur. Mrs. Benedict had one such crisis two days ago and it looks like she came out of it OK."

Mr. B. "But you agree with the plasmapheresis therapy?"

Dr. X. "Oh, absolutely. Apart from time, it's the only effective treatment we can offer these patients."

Mr. B. "If this was your wife, would you do anything different? Anything at all?"

Dr. X. "Mr. Benedict, if this was my wife I would not change a thing. And if my wife happened to be in this hospital, with this condition, I would not transfer her to New York. I would leave her right where she is."

* * *

That first week in MICU Naomi could only move her eyes, eyelids, and some facial muscles. She managed a weak smile but was unable to open her mouth wide or turn her head. Even so, Mr. Benedict decided the children should see their mother. His story about "infection" was wearing thin and the kids – a six-year-old boy and ten-year-old girl – began to wonder out loud if Mommy was dead.

I was ambivalent about the kids seeing their mother paralyzed. They last saw her the morning of hospitalization. A visit when she was still paralyzed could help or hurt her, and I wasn't sure which. I suggested to Julian that he seek Naomi's opinion and he agreed. To his question ("Naomi, do you want the kids to come here?") she vigorously moved her eyes up an down. Yes!

Arrangements were made for the afternoon of March 14, her seventh day of hospitalization. The night before the visit Julian tried to prepare the children, by explaining that Mommy was too weak to move and would not be able to talk, but that she loved them very much, and that she would come home faster if they let her know how much they loved and missed her.

About an hour before the appointed hour nurses tied Mrs. Benedict's hair back in a bun and attached a pretty red ribbon. They also applied a small amount of makeup to her lips and cheeks. With the head of her bed raised to 45 degrees Naomi was sitting almost upright. Her head was buoyed by a pillow and turned to the left, so that she faced the side where her kids would stand. The tracheostomy tube and connecting hoses were covered discreetly with a bed sheet. From a distance Mrs. Benedict looked almost normal, like someone sitting in bed watching television.

Shortly after 4 p.m. Julian and the kids arrived. Both children were smartly dressed in school clothes. By pre-arrangement Julian took them straight to her room without any introductions to me or the staff. Worried about another autonomic surge, I discreetly stood in one corner of the room where she could not see me. Julian and the kids entered and walked to her bedside.

"Hi honey," said Julian. "I have Kevin and Cynthia with me."

Naomi managed a weak smile, an idiot-like grin the kids had never seen before. I saw it from across the room and shivered. Kevin and Cynthia just stood there, staring at their mother. For a few seconds – it seemed like a few minutes – no one said anything. I wondered: Did we make a mistake, letting them see her like this?

"Hi mom," said Cynthia, the ten-year-old. Thin and pretty, she was destined to be a beauty like her mother. "Hurry up and get well. We sure miss you. Daddy's taking good care of us."

"Mommy, this is for you," Kevin said, and he showed her an 8 x 11 inch

picture he had drawn for the occasion. It displayed a red stick figure on a brown stick bed and big green block letters proclaiming GET WELL MOM SOON.

Mrs. Benedict wanted to smile and laugh and say what a wonderful picture it was, but all she could manage was the same feeble grin. Kevin didn't understand.

"Mommy, don't you like my picture? Mommy, why don't you hug me? I miss you Mommy!" He started to cry.

Cynthia nudged her brother and whispered sternly: "Kevin, Mommy can't hug you. She can't move right now."

The boy jumped away from Cynthia and tried to climb into his mother's bed. Julian pulled him back and he started to scream and cry louder. "MOMMY! MOMMY! MOMMY!"

Tears welled up in Naomi's eyes. Suddenly I felt awful. What must she feel? How unfair! Why had we let them come in? I wanted to leave and let Julian handle this visit in his own way but the cardiac monitor showed an accelerating heart rate. One hundred ten. One hundred twenty. One hundred sixty.

Although we needed to treat her quickly I spoke up without trying to sound excited or anxious. "Julian, I think we'll have to give her something. You better take them outside." He looked at me and I pointed to the cardiac monitor. "Let's go kids," he said, and ushered them out.

* * *

The next two weeks were dismal for the Benedicts. Lack of any clinical improvement was wearing Julian down. One telephone conversation after a plasmapheresis treatment on March 20:

"Hi Larry, any change?" (By then we were on a first name basis).

"Not yet Julian. How are the kids?"

"Oh, they're fine. No permanent damage. They're just waiting for Mom to come home."

"I know. This is difficult for them. Well, tell them she will be home. And tell yourself that, too. It just takes time for the nerves to regenerate. If she remains stable she should improve." Fortunately there were no more autonomic crises, no more emotional upheavals. Just the paralysis. Day after day of para-lysis.

Then a change. Twenty days and six plasmapheresis treatments after coming to the hospital Mrs. Benedict began to move her *right index finger*. Regeneration!

"You're getting better," I told her. "You really are." We noted another big change: a broad, deep smile, far from the idiot grin displayed when her kids visited. I called Julian to relay the good news.

"How long will it take her to fully recover?" he asked.

"I don't know. Dr. McAllister says it could still take months, but at least nerve regeneration has started."

Without exercise – even the passive variety – paralyzed muscles develop disuse atrophy. Even after the nerve tissue regenerate the muscles can remain severely weak even. Since day one we had provided range of motion exercises to Naomi's paralyzed limbs. Despite the exercises (and tube feedings) there was some loss of muscle mass and her weight was down twenty pounds at the end of three weeks. She needed more exercise. From the orthopedics department we arranged to borrow a mechanical exerciser, a machine that continually moves the leg or arm to help maintain muscle tone. The machine is used mainly to exercise limbs after orthopedic surgery but we have also found it useful in some paralyzed patients.

Naomi had full sensation in her extremities so we alerted her to possible pain. "This contraption will keep your muscles fit," I explained. "If it causes you any pain or discomfort blink your eyes rapidly."

We secured her right leg to the exerciser. First the machine extended her leg. Then flexed it. Extended, flexed. Extended, flexed. A full range of motion every 12 seconds. Fortunately, she felt no pain.

Naomi continued to improve and by the end of March she could move her arms and head, but could not write or hold a glass. Also, her breathing muscles were still very weak. Normally, we can generate enough muscle strength to suck up a column of water to a height of about 100 centimeters (39 inches). People with respiratory failure from muscle weakness can suck up no than 20 centimeters (8 inches). The first time we tested her respiratory muscle strength, three days after the tracheostomy, Naomi managed only 10 centimeters of 'sucking' pressure. Now it was up to 18 centimeters. Still low but increasing!

"You continue to improve," I said, "but we have to go a few more days before you can get off the machine. You'll make it. As soon as you do we're going to throw a big party. If it's OK with you we'll keep it private. No one from the media will be invited." She smiled the equivalent of a hearty laugh.

We kept at it. Range of motion exercises. Tests of breathing strength. Tube feedings. Constant monitoring. She had settled into MICU and was beginning to feel almost "at home." She knew all the nurses and felt comfortable with her surroundings. The trauma of her kids' visit was ancient history.

To keep her mind occupied Julian brought in cassette selections from Books on Tape, which she listened to through an ear plug. In two weeks she went through Mark Twain's *Life On The Mississippi*, *The Cardinal of the Kremlin* by Tom Clancy and *The Bonfire of the Vanities* by Tom Wolfe.

* * *

On April 12 her inspiratory force was 24 centimeters of water. We disconnected the ventilator and gave her supplemental oxygen through the tracheostomy tube. Now, for the first time in over a month she was breathing entirely on her own. Since her inspiratory muscle strength was still 'borderline' I put her back on the ventilator after two hours.

"You did just great," I said, "but I don't want you to tire out. It's best if we do this a little bit each day. Tomorrow you'll go for four hours off the ventilator."

She moved her lips: "Where's my party?"

Naomi improved almost as rapidly as she had crashed. The next day her inspiratory force was 30 centimeters. I took her off the ventilator with the understanding that she would be re-connected after four hours.

For three hours and forty-five minutes there was no sign of fatigue or respiratory distress. Encouraged, I told the nurses to leave her off the machine indefinitely, with the idea that she might go the whole night unassisted.

Fifteen minutes later Naomi asked to see me. She had recently regained some use of her writing hand and was now communicating with paper and pencil. She wrote: BACK ON THE MACHINE?, meaning the ventilator.

"How do you feel?"

TIRED

I checked her respiratory rate, inspiratory force, cardiac rhythm, and blood pressure. They all pointed to one fact: physiologically, she didn't *need* the ventilator. Psychologically was another matter. "Do you think you still need the breathing machine?" I asked.

YES

"Do you want to try and go without it a few more hours?"

NO

I have seen this response many times. Removing a patient from prolonged artificial ventilation often requires physiologic *and* psychologic adjustment. It makes sense. What has been critical life support is being discontinued; even though the patient no longer needs it, the "weaning" process takes time.

"OK," I said. "I think one more night. We'll put you back on the ventilator tonight. Tomorrow morning we'll disconnect it and let you go all day. If you do well during the day you'll be able to go all night without it. I promise." She liked the plan.

On April 14 she went all day without needing or asking for the ventilator. And all night. On April 15 we had our party.

Follow up

Naomi Benedict continued to recover muscle strength. She began swallowing food on April 18 and regained movement in her legs by April 21. On that date we removed her tracheostomy tube. On April 25 she was

transferred out of intensive care to the hospital's rehabilitation unit.

As expected she required extensive physical rehabilitation. On May 8, with the aid of a lightweight aluminum walker to keep balance, she took her first unassisted steps in two months. She went home from the hospital May 10.

Naomi continued muscle training exercises as an outpatient. Though progress was slow recovery was total. She started driving again in the middle of July and by September was back to a full schedule of meetings, parties, and charitable activities. She reports that Kevin and Cynthia are none the worse for her two month absence and that life is back to normal.

The only scar from her ordeal is physical and small, where the tracheostomy tube entered her neck. She usually covers it with a high collar blouse.

-- END –

21. The Wild Man

Joe Cartney was one of our most difficult patients, and we almost lost him. He showed up in our emergency department – dumped there is more accurate – one cold Saturday morning. It seems his brother or brother-in-law brought him in because he, Joe, was "taken real sick all of a sudden." After imparting this message to one of the secretaries, Joe's relative sat him on a waiting room chair and went to "park the car." He did not return.

Left behind was a young man too sick to stay in any chair. He fell to the hard linoleum floor and proceeded to moan and wail. The ED nursing staff responded with a quick check of his vital signs, and lots of questions: "Who is this? Where'd he come from? What's his name?" Only the hapless secretary had any information: "Mr. Joe Cartney...brother [?brother-in-law] brought him in...said he's real sick, then left...said he'll be right back...didn't leave any phone number...didn't sign any forms."

As for the patient, all he could do was scream and flail his arms. Four of the staff lifted Mr. Cartney to a stretcher and brought him to the treatment area, then placed him in a bed. If acutely ill, at least he was in the right place.

He could give no history worth a penny. He just moaned and screamed at the slightest provocation, calling for "Emma," presumably his wife or girlfriend, and various other people who he could/would not identify. He knew his name but not the day, year, where he was or how he got there. He could not answer any question coherently. He frequently resorted to scatological phrases like "you mother f-----." Joe Cartney was, in a word, delirious; though acutely ill, he was simply unable to communicate his problem. The ED staff had no more history than a veterinarian presented a sick dog found on the street. Methodically, the ED physician and nurse documented what they could:

Vital signs. Blood pressure stable at 130/70; pulse irregular at 150 to 170 beats/minute (normal is less than 100/minute). Temperature 101 degrees.

Physical exam. Young male, ?age. Medium build. Appears disheveled, unshaven, dirty. Skin moist and sweaty. No obvious signs of trauma or recent injury. Three tattoos noted: "Emma" over left arm, skull and cross bones over right arm, and "Born to die" over chest. Scars over abdomen, right thigh (?knife wound). No neurologic defects, nothing to suggest he had suffered a stroke. Able to move all his extremities.

Cardiac rhythm. Irregular, with many extra heart beats. Technically,

"supraventricular tachycardia with numerous multi focal beats."
Arterial blood gas test. Adequate oxygenation and ventilation, (i.e., his lungs were working normally).

Other blood tests. Results pending, including a "toxic screen" of his blood and urine for drugs commonly used to overdose.

Chest x-ray. Clear; no evidence for pneumonia.

Those first few minutes Mr. Cartney didn't need cardio-pulmonary resuscitation or anything else heroic. He did need lots of attention, to keep him stable and in bed, to exchange his clothes for a hospital gown, and to find out what was ailing him. First guesses were acute alcoholic intoxication or withdrawal from alcohol, toxicity from illicit street drugs like PCP (phencyclidine, or "angel dust"), or some type of nervous system infection, either meningitis or brain abscess.

He did not carry the smell of alcohol. Nor did he appear yellow, bloated or wasted like an end-stage alcoholic. As for PCP, detection requires a urine measurement, ordered as part of the tox screen. In cases like this, until all the information is available, it is best to assume some type of drug overdose and manage accordingly. On that assumption the ED physician placed a large, snake-like hollow tube through Mr. Cartney's nose and into his stomach. Through the tube she squirted some "activated charcoal," a highly absorbent black slurry that binds whatever pills or tablets it touches. After a few minutes the charcoal was sucked out of Mr. Cartney's stomach and with it, hopefully, any poisons he might have swallowed.

Of course the ED team had no specific history for drug overdose, so other possible diagnoses were also considered. Like meningitis. Meningitis can be confirmed (or excluded) with a spinal tap and quick exam of spinal fluid under a microscope. But a spinal tap requires careful insertion of a thin needle in the patient's lower back, and Mr. Cartney could not stay still for that procedure. For the same reason a brain CT scan was not feasible, since it also requires a still patient.

And still he was not. Within 20 minutes of arrival he was referred to as the "wild man." To keep him in the bed all four limbs were bound with leather restraints securely attached to the bed frame. Every minute or so he would try to sit up, pull against the restraints and bellow, "Get me out of here you mother f------!," or something similar. Joe Cartney was a danger to himself so he needed the restraints.

The ED's job is to treat emergencies and then triage. On the battlefield triage means deciding which soldiers to let die and which to try to save. In the ED triage means proper disposition of the patient – treat and release, or treat and admit to the appropriate part of the hospital: intensive care,

medical or surgical ward, etc. Since the ED is on ground level patients are either sent "upstairs" or discharged.

Acutely ill patients who pose a diagnostic dilemma, like Joe Cartney, may be kept downstairs for hours, while tests are run and the problem sorted out. Does the patient need surgery? Admission to an intensive care unit? More tests in the ED? Joe Cartney was the epitome of diagnostic dilemma, but the ED staff did not wait for more tests; they sent him up to medical intensive care after only 30 minutes in the ED, a record for a patient not needing emergency surgery.

I was already in the ICU, making Saturday morning rounds, when the call came from the ED nurse. One of the ICU nurses answered the phone, took the message and hung up the receiver while announcing, "We have a wild one coming. A male."

"What's wrong with him?"

"They don't know. Possible overdose, possible meningitis. They said he's wild and very sick."

Oh, oh.

* * *

Mr. Cartney arrives accompanied by three security officers, plus an ED nurse and physician. He is half sitting on the transport bed, bellowing and pulling on his restraints. A thick black tube sticks out of his nose and I see splotches of charcoal on his bed hospital gown.

The ED physician gives me a one minute capsule history, finalizing her recitation with, "This guy's real sick, we don't know why, but he needs to be in MICU. Maybe an overdose, maybe meningitis, we don't know. There's nothing more we can do for him downstairs. He's all yours now."

We undo Mr. Cartney's restraints and transfer him to the ICU bed, then re-attach the leather straps. The ED and security folks promptly disappear. As the MICU nurses go about their business, I move to the head of the bed and look at our new patient. He becomes eerily silent and stares back at me. He is maybe 29 or 30 years old. His eyes are wide open and he does not appear to be in pain or even any distress (then why all the wailing?). The sweat on his brow, dilated pupils, and pulsating neck veins all give the appearance of acute intoxication, but with what?

I try a direct approach. "Mr. Cartney, what did you take?"

No answer.

"Did you take PCP?"

"Yes," he responds, slowly and in a low moan. His answer is not convincing, so I persist.

"Did you take alka seltzer?"

"Yes."

"Did you take XYZ?"

"Yes."

We seldom ask yes or no questions because you can't trust the answers, but this guy is not coherent and I thought it worth a try. Alas, he is not going to give any meaningful medical history.

Suddenly he grimaces and starts yelling again. "EMMMMA!"

Mr. Cartney, who's Emma?"

He tries to sit up and only succeeds in pulling on his restraints. "Let me out of here!" he bellows, then collapses back to the bed.

We take his vital signs. Blood pressure 120/65, pulse irregular and ranging from 150 to 180 beats per minute, temperature 100.5. His electrocardiogram shows chaotic supraventricular rhythm, not immediately life threatening but definitely, severely, abnormal. Throughout all this activity I, we, everyone, wonders – what's wrong with him? What's the diagnosis here?

In another ten minutes we have more data. His tox screen is ...negative. Neither the urine nor blood samples reveal any of several drugs commonly taken in overdose. Some drugs are looked for only in the urine, others only in the blood (serum), and a few in both body fluids. Here is Mr. Cartney's toxic screen report:

*** Drug Overdose Screen, Urine***
URINE DRUG OVERDOSE SCREEN IS NEGATIVE. URINE DRUG OVERDOSE SCREEN INCLUDES TESTING FOR:

AMITRIPTYLINE, AMPHETAMINE, CIMETIDINE, COCAINE, CODEINE, DESIPRAMINE, DIPHENHYDRAMINE, FENTANYL, FLUPHENAZINE, HYDROMORPHONE, IMIPRAMINE, LIDOCAINE, MAPROTILINE, MEPERIDINE, METHADONE, METHAMPHETAMINE, METHYLPHENIDATE, MORPHINE, OXAPINE, OXYCODONE, PENTAZOCINE, PHENCYCLIDINE [PCP], PHENTERMINE, PHENYLPROPANOLAMINE, PROPRANOLOL, PROPOXYPHENE, PSEUDOEPHEDRINE, PYRILAMINE, THIOTHIXENE, TRIPELENNAMINE

*** Drug Overdose Screen, Serum***
SERUM DRUG OVERDOSE SCREEN IS NEGATIVE. SERUM DRUG OVERDOSE SCREEN INCLUDES TESTING FOR:

ALPRAZOLAM, AMITRIPTYLINE, AMOBARBITAL, AMOXAPINE, AMPHETAMINE, CARBAMAZEPINE, CHLORDIAZEPOXIDE, CIMETIDINE, CYCLOBENZAPRINE, DESIPRAMINE, DIAZEPAM, DOXEPIN, FLURAZEPAM, GLUTETHIMIDE, IMIPRAMINE, LIDOCAINE, LORAZEPAM, MAPROTILINE, MEPERIDINE, MESORIDAZINE, METHAQUALONE, NORTRIPTYLINE, PHENOBARBITAL, PHENYTOIN, PRAZEPAM, PYRILAMINE, QUININE, SECOBARBITAL, TEMAZEPAM, THIORIDAZINE.

So PCP toxicity is, apparently, ruled out. Most of the other drugs tested for are analgesics, tranquilizers, sedatives and anti-epileptics. Also looked for are cimetidine (brand name Tagamet, a commonly-prescribed anti-ulcer drug) and propranolol (brand name Inderal, used to treat various conditions including high blood pressure and migraine headache). Obviously, not all drugs are tested in these two tox screens, but we must go with the information available. So far, we have no evidence for drug overdose.

Now we must make sure he doesn't have a treatable problem like infection (as opposed to an untreatable one, where you just support the patient and hope he gets better). As for his cardiac arrhythmia, it is clearly the result of some disorder and not the cause of his problems. His blood pressure is normal and he is not in shock. It is his fever and confusion that make meningitis and brain abscess a major concern.

Meningitis, infection of the covering of the brain and spinal cord, is a medical emergency. It can kill quickly, particularly when caused by some types of bacteria. Brain abscess is a pocket of pus inside the brain and also a very serious problem; without surgical drainage of the pus the patient will succumb. Mr. Cartney was given a broad spectrum antibiotic in the ED in case he does have meningitis, but good practice mandates a spinal tap so the spinal fluid can be cultured for infecting organisms. There are too many different causes of meningitis to rely on empiric antibiotic therapy.

But we have the same problem as the ED staff. Mr. Cartney cannot be safely positioned for a needle in his back. He is simply too combative. And his heart beat is chaotic, a concern even though he has a decent blood pressure. We are stumped. What to do?

More test results return. His urine analysis is normal, no evidence for infection or diabetes. Kidney and liver function tests are normal except for a low serum potassium, a key electrolyte in the body's cells. Why a low potassium? Did he overdose on some diuretic medication that depletes the body of potassium?

And where is his family? Here is this fabulously ill guy, now in the hospital over 90 minutes, and no one is around who knows anything about him. Where does he live? With whom? Who brought him in?

The information in the hospital's hastily-put-together chart is meaningless. Phone number 999-9999. Social security number 000-0000-000. (Who programs these computers anyway? Why not just "unknown" instead of a funny number?)

We call in a cardiology consultant. Dr. Sally Zingale is next door making rounds in the coronary care unit, and she comes right over. Yes, she says, the heart rhythm is chaotic. A weird pattern of atrial tachycardia and atrial premature beats. And no, there is no specific treatment, at least not right now. She specifically does not recommend any anti-arrhythmic medication. "Find the underlying cause," she says. "I'll come back shortly."

"Thanks."

* * *

So here is the situation. This guy is sick, sicker by far than any other patient at the moment, and we have no idea why, and no medical history. We suspect a drug overdose or nervous system infection. The tox screen is negative and he is too combative to do a spinal tap. What next?

Do not think this problem is handled in isolation. At any given moment I am consulting with two or three other physicians, including residents in training, plus the patient's ICU nurse and anyone else who will listen. It is a weekend so there aren't that many people around, but I have help. What I don't have is a diagnosis.

Joyce Munson, one of the nurses, wonders: "Dr. Martin, what about rabies?"

"Rabies?"

"Yes, don't people with rabies act like this guy?"

"You mean wild and delusional?"

"Yes, just like our Mr. Cartney."

"Have you ever seen a case of rabies?"

"No, but I've seen it in the movies."

Rabies is a viral infection following the bite of a rabid animal (hence the term), usually a dog or bat. The virus attacks the nervous system and, once symptoms begin, death is inevitable. There is no treatment and no cure. A series of vaccinations can be given to prevent spread of infection, but the shots must be started right after the animal's bite, before any symptoms begin.

Rabies is an interesting idea, but an unlikely diagnosis. No one in our hospital has ever seen a case of rabies. In fact there has been no human case in our state in many years.

"Could be, Joyce, but I seriously doubt it. It would be the first case ever in this hospital, and it seems so improbable. From what I've read, rabies victims maintain muscle spasms constantly, especially the facial muscles. Mr. Cartney's grimacing comes and goes. He looks more like some type of drug toxicity. In any case, we've got to look for something treatable and rabies isn't treatable. But if we do the spinal tap I'll save some fluid and check for the rabies virus if nothing else turns up.

"OK. Just a thought," she says.

Basically, I see two options. Watch him in his current condition, see what happens over the next 24 hours and wait for some more test results (of thyroid function, for example; extremely over-active thyroid is a remote consideration). If some drug toxin in his body is being slowly excreted, he might improve over time. But if we miss a treatable infection, he could die for lack of the right antibiotics.

Alternatively, we can sedate and paralyze him with medication, then do

a spinal tap and CT scan. This course of action will require connecting him to a breathing machine, since drug-induced paralysis will stop all spontaneous breathing.

Death from meningitis for want of a spinal tap and proper fluid cultures is probably indefensible. With sedation we can control him, do the spinal tap, and maybe even figure out why his heart rhythm is so abnormal. On the other hand, intubation and artificial ventilation are not without hazard, and if there is a complication...

Jed Warner is with me, a top flight medical resident. I lean heavily on his judgment.

"Jed, what do you think we should do?"

"Knock him out and ventilate him." Jed is a direct kind of guy.

"Just like that? But his breathing is OK. Don't you think that's a bit drastic? I mean, he's a little wild, but he doesn't require artificial ventilation for any of the usual reasons. His lungs are normal."

"I agree, Dr. Martin, but you're not going to get anything done with him. We can't do a spinal tap or CT scan. You can't even check a blood pressure measurement without full leather restraints."

"What if we have trouble intubating him? What if we paralyze him and the tube comes out of his throat (these things have happened). What if..."

"Obviously anything can happen, Dr. Martin, but I don't know where we're going to go with this guy. He could improve, but he's got a low potassium, a chaotic rhythm, and he could also get worse. With either meningitis or some poison in his body he could stop breathing on his own. Then where are we?"

Jed is right. I just needed a little prodding to help make the right decision. We proceed to paralyze and intubate Mr. Cartney. We ask the nurse to inject 2 mg of Versed (a sedating drug), followed by 2 mg of Pavulon (a paralyzing drug). All the while we monitor his heart rhythm. When Mr. Cartney is sufficiently relaxed Jed proceeds with the intubation, which requires placing a foot-long plastic tube into Mr. Cartney's trachea. Jed has experience with this procedure.

* * *

As Jed struggles with inserting the narrow tube into Mr. Cartney's trachea, there is a sudden drop in blood pressure. The reason is readily apparent from the cardiac monitor.

Someone yells: "He's in vee fib!"

I stare at the monitor in utter dismay.

"Let's defibrillate. Is he being ventilated?"

"Yes, the tube's now in place."

"Stand back. Get ready."

Mr. Cartney now has the most chaotic of all cardiac rhythms, ventricular

fibrillation. It is the rhythm of sudden death. We have changed a chaotic *atrial* arrhythmia – one that provides a decent blood pressure – into a chaotic *ventricular* arrhythmia. Ventricular fibrillation causes the heart's major pumping chambers (the ventricles) to undulate like a bag of worms. A heart in "vee fib" cannot pump any blood to the body. Uncorrected, ventricular fibrillation leads to brain death in about four minutes.

(Ventricular fibrillation is the most common cardiac mechanism leading to "sudden death." If you saw the movie Jurassic Park, remember the boy jolted by electric shock? Ventricular fibrillation.)

Joyce presses the buttons on the two defibrillator paddles straddling Mr. Cartney's chest. There is an audible "click" as 200 joules of energy surge through his heart. And I am thinking. 'Damn. Damn. Damn. How did this happen? Are we going to lose him? Oh, damn. Damn, damn, damn. Please don't die.' This is actually what I am thinking.

"OK, he's back in a supraventricular rhythm. I see narrow complexes. Do we have a blood pressure?"

"One twenty by palpation."

"Start lidocaine. Let's give him a hundred milligrams."

Another minute passes. We all stare at the monitor.

"Jesus Christ! He's in vee fib again!"

"Shock him."

"How much?"

"The full 350."

"Stand back."

He is defibrillated again, this time at the maximum energy level, 350 joules. Meanwhile, one of the technicians continues to ventilate him through the endotracheal tube with an AMBU bag. Things are too unstable to rely on a mechanical ventilator right now.

"OK, he's back in some sort of regular rhythm. Call cardiology again." A minute passes and Dr. Zingale is back in the room. We hand her a bunch of long rhythm strips from the cardiac monitor. They document the sequence of Mr. Cartney's alive-dead-alive-dead-alive cardiac rhythm.

"Vee fib," she says.

"I know, Sally. We shocked him out of it twice. I just gave him a bolus of lidocaine. Any other suggestions?"

"Do you have a blood gas and electrolytes?"

"They're cooking."

"Let's start him on a lidocaine drip at two milligrams a minute."

"OK."

Arterial blood gas results return and actually show decent oxygen and carbon dioxide levels. And his potassium is now normal (he has received potassium replacement in the IV fluids).

Ventricular fibrillation does not return. We connect his endotracheal tube to the breathing machine. By now he is quiet, paralyzed from

medication, and his heart is back to its previous chaotic (but life-sustaining) atrial arrhythmia.

We wait a half hour to make sure he is stable, then do the spinal tap. The procedure goes without a hitch. The fluid is clear. No sign of spinal infection.

"Let's get him down for the CT scan."

A cadre of four people wheel Mr. Cartney and his machines to the CT scan suite in the basement. A half hour later we have the scan results. Normal. No brain abscess.

* * *

It is now two in the afternoon, some six hours after Mr. Cartney came to our ED. He is back in his room in the ICU and I have long since given up my afternoon plans. I sit staring at a mass of lab data, and at his cardiac monitor. The answer is staring back at me but I don't see it. It is too obvious, I suppose, but there it is. Chaotic heart rhythm, delirious patient, low potassium level, no apparent infection. What did this man take that we haven't measured or looked for? I work with this substance all the time, yet inexplicably do not think of it until...

"Dr. Martin, his family's in the waiting room."

"What? Mr. Cartney's family?"

"Yes, they want to know if they can see him."

"See him? You bet. I'm going out to talk to them."

Jed and I amble out to the waiting room. I am more curious than I can remember to meet a patient's family.

In the waiting room are two men and a woman. The men, like Mr. Cartney, appear dirty and disheveled. They have grease in their hair, on their clothes. Perhaps, I think, they just finished working in a gas station. And they smell of cigarettes. But their appearance is not important. I need *information*.

I introduce myself and Jed. One of the men speaks.

"This here is Joe's wife, Emma. How's he doing?"

The woman appears to be about 25. I notice she wears no makeup. She has that depression era look: pallid face, faded dress, work style shoes. She seems both anxious and depressed at the same time. I immediately feel sorry for her, and not just because her husband is in a coma.

"He's alive, but we have no idea what's wrong with him. Who brought him in."

"I did," replies the same man.

"Are you his brother? Why didn't you stay to give more information?"

"No. We're Emma's brothers," he says, and points to the other man, who doesn't speak but gives a polite nod. "My name's Billy Echol. I brought him in. I was double parked, so I told the girl I got to go get

Emma."

"Why didn't you return right away? We really have no idea what's happened to Mr. Cartney. Why didn't you at least call us?" I don't know why I'm giving this guy the third degree, but I am annoyed and can't help myself. My questioning doesn't faze him.

"No phone, doc. And we had some car trouble. It's fixed now."

I am poised to ask numerous questions about Mr. Cartney. When did he get sick, what were his initial symptoms, is there any history of animal bites, any history of drug overdose, do they know if he took any pills, and so forth and so on. I am expecting a difficult-to-get history with many blind alleys. But I am wrong. His brother-in-law heads me off.

"We found this in his room." He hands me a note.

A note from Joe Cartney. *A suicide note!* In block letters and crudely written on an envelope:

EMMA. CANT GO ON ANYMOR.
BETTER WITH ME GON.
LOV, JOE

"Then he took something!" I am excited. We are getting close to an answer. "What did he take? Do you know what he took?"

Billy has that information also. He hands me a plastic pill bottle. "This here's Emma's asthma pills. It's empty. She says it was full yestiday."

I stare at the label. It is the missing piece to this puzzle.

```
Emma Cartney.
Theodur 300 mg, # 60
Take 1 tablet twice
a day with meals
```

Theodur. *Theophylline*. Of course! It all fits! He overdosed on theophylline, a common asthma drug.

"Jed, let's get a stat theophylline level on Mr. Cartney. Call the lab and tell them to please run it right away. After that, take Mrs. Cartney and her brothers in to see him. I'm going to call the kidney dialysis service."

I return to the ICU and place a call to Dr. Richard Malcolm, the physician on weekend call for the kidney dialysis service. If Mr. Cartney took 60 tablets of Theodur he may need hemoperfusion, a sophisticated technique that can wash out theophylline from the body by passing the patient's blood over a special column of charcoal. The charcoal column absorbs theophylline and the cleansed blood is then returned to the patient. It is a sophisticated and rarely performed procedure. Most overdose patients don't need hemoperfusion, and it is only effective for some types of drugs.

Theophylline is one of them.

Theophylline, of course, is widely used to treat asthma. The Physician's Desk Reference of prescription drugs lists over two dozen oral theophylline preparations, all for asthma and asthma-related conditions. Most are theophylline-only drugs but a few contain theophylline in combination with another asthma-treating drug, ephedrine. Any of the theophylline preparations, taken in excess, can cause severe theophylline toxicity. Here is a partial list, in alphabetical order by brand name.

Aerolate
Asbron
Choledyl
Dilor
Marax
Quadrinal
Quibron
Respbid
Tedral
Slobid
Theo-24
Theodur
Theolair
Theophylline (generic)
Uniphyl

Mild toxicity from theophylline is actually fairly common. It happens to patients who take the drug as prescribed but, for one reason or another, build up a higher-than-expected level; just stopping the drug is all the treatment needed.

Even people who intentionally overdose with a theophylline preparation rarely need hemoperfusion. Usually the drug dissipates naturally in the body and clinical improvement rapidly follows. But there is a limit to how much theophylline the body can tolerate. Mr. Cartney's case turns out to be the mother of all theophylline overdoses.

* * *

"I can't do hemoperfusion without a drug level."

"Richard, the theophylline level's being run now. This guy almost died twice on us already. The whole thing fits. Potassium goes down in theophylline overdoses. His cardiac disturbance is classic for theophylline toxicity. And his delirium fits the picture also."

"Why didn't you draw the level earlier?"

"We didn't think of it. It's not part of the routine tox screen and we had

absolutely no history. We're all smart in retrospect."

"When will it [the drug level] be back?" It is now 2:30.

"About three o'clock, the lab said."

"Well, we're ready to go," Dr. Malcolm says. "My technician is warming up the equipment. If it is theophylline, how high do you think his level is? Do we know when he took the tablets?"

"No, his wife found him puking this morning. Her brother lives next door. He rushed him over here, but gave absolutely no information. I guess no one did any looking around the house at first. Anyway, his brother-in-law just dropped him off and left. Then they found the note and the pill bottle. The way he's behaving, I'd say he swallowed the whole bottle about midnight. The pills were probably all dissolved into his blood by the time they pumped out his stomach this morning."

"So what do you think his level is?" Dr. Malcolm asked. The question was like, "So what do you think the final score will be?" I took up the challenge, and called over to Jed, who was writing some notes.

"Hey, Jed, what do you think Mr. Cartney's theophylline level is?"

The therapeutic theophylline level – the amount in the blood that is suppose to help asthma patients when they take the pills as prescribed – is between 5 and 15 milligrams per liter, written '5-15 mg/L'. Patients tend to get sick with nausea, diarrhea, or palpitations when the level exceeds 20. Above 30, and a patient will likely be admitted to the hospital; above 40, to the intensive care unit. Most experts recommend hemoperfusion when the level exceeds 80. The highest level I have personally seen to this point is 84, and that patient had seizures.

"Oh," said Jed, "considering they've already pumped out his stomach, I'd say about sixty."

"OK, let's write it down. Jed says sixty. I say eighty-five." I write both numbers on a piece of paper and put our initials by them. "Now your turn, Richard."

"Oh, I don't know. You see more theophylline toxicity than I do. We did hemoperfusion on a guy last year with a level of eighty-four [the same patient mentioned above]. He was pretty sick too, and he had seizures. I'll go way out and say ninety for this guy."

I write his guess down with the other two. Our range is 60-90. Whoever is closest, it is clear we all suspect a very high level.

But not high enough.

"Doctor Martin, the lab's on the phone." I take the receiver.

"Yes?"

The lab technician is excited. He asks if the patient is still alive.

"Yes. Are you sure about the level?"

He's run it twice, and is sure. I thank him and hang up the phone.

"Wellllll?" Dr. Malcolm asks, "the winner is..."

"One hundred and sixty seven."

"Wow!" This from Jed.

* * *

The hemoperfusion goes smoothly, as Mr. Cartney's blood is circulated through a machine that contains the special charcoal column. Blood is routed from the large femoral vein in his thigh, bathes the charcoal column so theophylline can be absorbed, and is returned to his body through the same vein. Quite an elegant process. We remove Mr. Cartney's stomach tube as it is now superfluous.

After four hours of hemoperfusion his theophylline level is down to 88 mg/L. After eight hours it is 56. At this level his heart rhythm miraculously returns to normal. We stop the sedative medication. He is beginning to wake up.

Twenty four hours later his theophylline level is 21 and he is fully awake. We disconnect the ventilator and pull out his endotracheal tube. He seems all better.

And he is a nice guy. No more wild gyrations. No more foul language. He seems both pleasant and contrite.

"What happened?" I ask.

"I don't know."

"Did you want to kill yourself?"

"Not really. We jist had some problems. I'm sorry now. I ain't going to do it again."

"How many pills did you take?"

"I don't know. The whole bottle, I guess. They was Emma's pills."

"You know you almost died?"

"I'm glad I didn't."

"We have to ask a psychiatrist to see you."

"OK."

Emma and two other family members come in next. Everyone is smiling, happy. We are all greatly relieved.

Mr. Cartney has no health insurance. The psychiatrist finds him to be clinically depressed, mainly over poor finances, lack of a job, and related woes. The entire family is on food stamps. He recommends that Mr. Cartney go to a county mental health facility. He does not believe Mr. Cartney is in danger of hurting himself again, at least not any time soon.

The next day, three days after his arrival, we discharge Mr. Cartney to the recommended facility; while there he receives some brief outpatient counseling, is deemed not suicidal by the county's psychiatrist, and is sent home.

* * *

A week later I present Mr. Cartney's case at a medical conference. Highlights:

- Most cases of theophylline toxicity are unintentional, usually because patients take (or are prescribed) more theophylline than they can handle.
- The first case report of hemoperfusion for theophylline toxicity dates to 1978. The patient, a 50-year-old woman, was taking Theodur for asthma while in the hospital. She developed refractory seizures when her theophylline level went to 46 mg/L. After 4 hours of hemoperfusion her theophylline level was down to 15.4 and her seizures gradually abated. However, she died two weeks later, of pneumonia.
- The highest theophylline level in any of the articles I reviewed is 210 mg/L. That patient died.
- Theophylline toxicity is the most common poisoning for which charcoal hemoperfusion is performed.
- The majority of patients with a theophylline level greater than 120 mg/L developed seizures, and over 50% of them died. It is unusual that with so high a level (167 mg/L) Mr. Cartney never showed any seizure activity.

* * *

About three months later, while making rounds in MICU, I am paged to the phone by Jed.

"Guess what, Dr. Martin. Mr. Joe Cartney, remember him? He was in the ED last night."

"Of course I remember him. Not another overdose?" I suddenly realize this cannot be, or I would have heard about it in MICU.

"No. No. He has a mild respiratory infection and just came in to get some antibiotics. He's actually doing OK. No major problems. I found out from one of the ED residents. Just thought you'd like the follow up."

"Thanks, Jed."

Not bad for a man who almost died. Twice.

– END –

22. ‘Lou Gehrig’ Strikes Again

I had long anticipated a distress call from someone in the Miraly family. When it comes I am at home, sleeping.

“Dr. Martin, Mt. Sinai Hospital calling.”

“Yes?”

“I have a Mr. Ernest Miraly on the phone. He says he needs to speak with you. Can I put him through?”

“Yes, go ahead.”

Mr. Miraly has a hoarse, quiet voice. Even face to face, during many sessions with the Miralys, I sometimes had difficulty understanding him unless I watched his lips move.

“Please speak up, Mr. Miraly. I can hardly hear you.”

“Dr. Martin, my wife Angela, she’s having trouble.”

“What kind of trouble? What’s happening?

“With her breathing. I don’t know what to do.”

“Is she awake? Can she talk?”

“No. I just called 911.”

“You called 911? How long ago?”

“Just before I called you. What should I do?”

He has already done it. The emergency medical squad will be at their house in a few minutes. And then what? Surely Mr. Miraly has understood my explanations, my recommendations, these past several weeks. I have done everything possible to warn him of this inevitable crisis, but he is not prepared for the sheer horror of watching his wife die.

“Mr. Miraly, please make sure the ambulance takes her to Mt. Sinai Hospital. I’ll meet you in the emergency department.” (EMS will get there sooner.)

“OK, Dr. Martin. You’ll call and tell them we’re coming?”

“Yes.”

“Will she go back into intensive care?”

“That’s up to you. We’ll see.” Then, as an afterthought I add, “And it’s up to her.”

I place a call to the ED and speak with the doctor on duty, Roger Stanton. I explain the case, tell him what to expect, and ask that they hold off intubating my patient, if feasible, at least until I get there.

* * *

Driving in I ruminate about Angela Miraly, and how she came to be my

patient. She was admitted to our intensive care unit only six weeks before, a transfer from the neurology service. About a month before that she had developed her first symptoms, an unsteadiness of gait, a slight slurring of speech, some weakness in her arms.

These symptoms grew worse to the point she no longer could drive. She saw her doctor, who sent her immediately to one of our staff neurologists, Edgar Mason. Edgar determined the likely diagnosis after his office exam, and recommended she come into the hospital.

Edgar had found telltale "fasciculations," fine, involuntary tremors of her arm and facial muscles. Progressive muscle weakness and fasciculations suggest a singular and terrifying disease, amyotrophic lateral sclerosis. In ALS there is atrophy and scarring of the spinal column nerves that control voluntary muscles. This nerve degeneration, of entirely unknown cause, can affect all the body's skeletal muscles, the muscles we can control at will (hence "voluntary"). Involuntary muscles, such as found in the heart and other internal organs, are not affected by ALS.

Could she have something else? Muscle weakness by itself suggested a dozen different diagnoses, from toxic drug ingestion to occult cancer to chronic fatigue syndrome. But she had no history of serious illnesses, no travel to exotic places, no drug use of any kind except post-menopausal estrogen, no history of depression, no evidence for cancer. As noted in her chart by the admitting resident: "First Mt. Sinai Hospital admission for this 57-year-old white female, who was healthy and well one month prior to admission when she first noted weakness in her arms . . ."

Healthy and well. Always on the thin side, about 110 lbs. and 5'4", a non-smoker, non-drinker, never before hospitalized except for childbirth. Mother of two grown children, grandmother of four, Mrs. Miraly was in her middle-age prime when ALS struck. Until weakness deprived her, she spent most of her free time in church work and other charitable activities. Her 62-year-old husband, a semi-retired accountant, was the other half of what, to any outside observer, appeared to be a long and satisfactory marriage. In all respects the Miralys were a pair of solid citizens to whom self-abuse was as alien as infidelity, a situation that made her disease seem so undeserved (compared, say, to cirrhosis in an alcoholic or emphysema in a smoker.)

Dr. Mason's plan was to do nerve conduction and other tests, confirm the diagnosis, and counsel the family. But on the second hospital day Mrs. Miraly choked on part of her lunch which, because of weakened swallowing muscles and a depressed cough reflex, went into her lungs. Because of weakened diaphragms (the principal muscles of breathing) she quickly developed difficulty breathing. That's when they sent her over to intensive care. On arrival to the ICU (about an hour after the aspiration episode) she was turning blue from lack of oxygen, so we intubated her trachea and connected the endotracheal tube to a mechanical ventilator. Then we sucked food out of her lungs and gave antibiotics. It took us about three days before

we could remove the tube and take her "off" the breathing machine.

By that time Dr. Mason had confirmed the diagnosis as "unequivocal ALS." He noted that her ALS was running a particularly progressive course, as opposed to a slower, more indolent course that afflicts some victims. He asked that I take over her care as principal physician since "her problems will no doubt be mainly respiratory." He would continue to "consult as necessary." What about treatment? Anything new? No, he said, nothing. Nothing new, nothing old. (The U.S. Food and Drug Administration has never approved a drug for treatment of ALS.)

So I inherited Mrs. Miraly's case by virtue of the disease's affect on her breathing, which remained fragile. Pulmonary function tests two days after extubation showed very diminished lung capacity, solely a result of weakened respiratory muscles.

I also took on the task of explaining what euphemistically might be called "the situation." This was not pleasant because ALS is inevitably downhill. It is progressive, no one ever recovers, and most people die within three years of diagnosis. Some patients can live longer with machine support (for breathing) and continuous nursing care (for feeding, bathing, cleaning, turning). It is a pure muscle-nerve disease, sparing all sensory and mental functions. Fortunately, ALS is not common; each year it is diagnosed in about one out of every 100,000 people.

ALS is popularly known as Lou Gehrig's Disease because the great Yankee first baseman's affliction with the disease occurred at the height of his career, and was widely publicized. ALS attacked him when only 35, at the start of the 1939 baseball season and his 17th year in the majors. At first, no one knew what was wrong. Despite increasing dis-coordination and difficulty ambulating, he suited up for the early games that season. But he could not hit or field the ball. Was the record-holding Gehrig (for hitting and consecutive games played) in a slump? Was he goofing off? What was going on? In May he quit professional baseball and traveled to the Mayo Clinic, where the diagnosis was made. At Gehrig's request, his doctor wrote a note for public release, explaining the diagnosis of ALS and why Gehrig would not be returning to the game. Gehrig became an instant hero. The Yankees retired his uniform number (4) and honored him at a special Yankee Stadium celebration in July. Gehrig died less than two years later, on June 2, 1941. No progress in ALS has been made since then. I had no good news for the Miralys.

Well, almost none. People *do* live with the disease, under circumstances not available in Gehrig's day. Perhaps the most famous living ALS victim is the British theoretical physicist Stephen Hawking (born 1942), who communicates via a computer and moves around in a motorized wheelchair. He can neither talk nor walk. Dr. Hawking was diagnosed with ALS many years ago, while studying for his doctorate. He has since formulated a major cosmological theory and authored best-selling books (*A Brief History of*

Time; *Black Holes and Baby Universes*). Despite almost total paralysis, Dr. Hawking remains an active participant in theoretical science, but his brilliance and motivation are far above that of the typical ALS patient. All the ALS patients I know have died within a few years of diagnosis.

While still in the ICU, but after extubation when she could again talk, I attempted to explain the diagnosis and all its ramifications. I did this at the bedside because I wanted her to be involved as much as possible in decision making. Since she tended to turn her head to the left (a result of some neck muscle weakness) I stood on her left side, next to Mr. Miraly. He stroked her hair while I spoke. She looked gaunt, older than 57, her hair white and stringy, cheeks sunken in, eyes wide open. It was a look, I thought, of fear and apprehension. It would have been easy to forgo discussing her future (I sensed she would never ask questions on her own), but I felt compelled to raise the issue as soon as she could respond. Events that had precipitated her ICU transfer could happen again, at any time.

"ALS is a progressive condition, Mrs. Miraly. Dr. Mason tells me it has already progressed quite a bit since your first symptoms a month ago. There may come a time when you can't breathe at all, without the breathing tube and a machine."

"When will that be, doctor?" she asked, innocently.

"No, dear," Mr. Miraly chimed in, "that's not going to happen, is it doctor?"

What? He didn't want to hear this.

"Well, it could very well, Mr. Miraly. ALS is a progressive condition, and unfortunately we can expect her muscles will get weaker over time and . . ."

He shook his head as if, perhaps, I was lying, or talking about some other patient. He wanted me to shut up and, for the moment, I did.

A day later she was out of intensive care, on a regular medical ward. I talked with them further. By this time she was eating again, but only puree foods since she couldn't handle solids well at all. Mr. Miraly was constantly at her bedside.

"We should talk about what can happen with this condition," I said. "There could come a time when you won't be able to eat on your own, or breathe without a machine. When that happens we can place a feeding tube in your stomach, and a breathing tube in your throat, like was done a few days ago. We can do these things for you, but that's how you will live. I don't think Mr. Miraly can care for you then, and it will mean going to some sort of long term facility, a skilled nursing home."

I tried my best to be direct and unequivocal, but also kind. I wanted to explain and make myself available for anything they wished to know. (Doctors can, with body language alone, make it clear they don't have time for questions. When that happens, patients usually don't ask any.) I sat down, looked in her eyes as I spoke, and made no sign of wanting to leave

the room.

I brought up the subject of "advance directives," decisions about what to do when the end comes. Would she want to be kept alive on a breathing machine? Some ALS patients opt for this. It can even be done at home, but requires round-the-clock nursing care. Others have asked to be let alone, to die naturally, and this can be handled at home as well (for example, through hospice care).

In patients with any progressive disease, or who may be terminally ill, we try to find out what they want (in terms of life support) when the end comes, and abide by their wishes if appropriate to do so. This patient-centered approach can help avoid 'medicine by default,' such as intubation when it's not what the patient wants. Does the patient really wish to be kept alive connected to a breathing machine, bed-confined, artificially fed liquid nutrient, day after day after day? In the world of ALS patients, Stephen Hawking is an aberration. Most of the time (always, in my experience) the patient ends up bed-confined, pitiable, merely *existing*. And a financial and physical burden to others, particularly caring and devoted family. I have sensed the family's collective sigh of relief when an ALS patient, bed-confined and physically wasted, *finally* dies.

But some people, given the choice, opt for machine living over the alternative, which is early death. And in the current *culture* of U.S. medicine – an amalgam of technology and litigation, of ethics and economics, of philosophy and common sense – the individual doctor is obliged to ask the dying patient: "What do *you* want?"

We've had the situation where ALS patients were intubated emergently and who, when it became clear they couldn't be taken off the machine because of respiratory muscle weakness, have asked us to pull out the breathing tube and let them die. Pull the tube out? It's an acceptable practice ethically, but stopping life support, especially in a mentally alert patient, engenders major stress for practically everyone. Of course the patient can be sedated and made unaware, in effect relieved of all anxiety; not so the doctors, nurses, or the patient's family.

Ethicists can (and do) argue that withholding and withdrawing life support are morally identical actions, but *they* are not the ones at the bedside pulling out tubes. Morality aside, it is certainly easier never to put the tube in than to have to pull it out. Don't put it in and the patient dies a natural death caused by no human hand. Pull it out and the patient dies because you pulled it out.

But if the patient is terminally ill she has the right to demand termination of life support. She can even demand it up front, using an advance directive. The most common advance directive is the living will, a legal document wherein you state what is wanted should you end up in a "terminal condition" or a "permanently vegetative state." The latter condition is rare, but "terminal condition" is not; it applies to a broad canvas

of disease states. Like beauty, "terminal" is in the eye of the beholder, a semantic conundrum that is the principal flaw of any living will. To my mind Mrs. Miraly was terminal because she had an incurable, progressively downhill disease; she would die of ALS much sooner than predicted from normal life expectancy, without *or* with machines. With machines and the enormous support they would mandate, she could live perhaps a few miserable years. Without machines perhaps a few months, or less. We were lucky to get her extubated and off the ventilator this time; her degree of muscle weakness made it clear that the next episode of respiratory failure would commit her to machines *forever*.

All this I explained to the Miralys, and told them about living wills. Over 10 days in the hospital I talked with them perhaps a total of six hours. I also met with their son and daughter, who came from out of town and stayed several days. Having families of their own, they had to return home, but by that time they and their parents heard much about ALS and what to expect.

Son and daughter agreed that their mother probably wouldn't want to be kept alive on a machine, but that "it was up to her." Mrs. Miraly never made any declarative statement herself. Mr. Miraly, for his part, would only listen politely. He told me they would think about the issue. In truth, they (patient and husband) simply didn't want to commit one way or the other. We arranged for a hospital bed at home, plus nursing care during the day and a special diet of soft pureed foods, and discharged her.

In the next six weeks I received three phone calls from their home, one from Mr. Miraly and two from her visiting nurse. A week after discharge the nurse called to get an antibiotic prescription, since Mrs. Miraly had developed a fever and possible bronchitis. I suspected pneumonia, but she didn't want to come to the hospital for a chest x-ray. The nurse's second call, a few days later, informed me that she was better with the antibiotic. I asked to talk to Mrs. Miraly, but when she came to the phone her speech was garbled. She was not in any distress, but weakness of the speaking muscles had made her unintelligible, at least on the phone. She had deteriorated further.

Mr. Miraly also called once to report on her condition, as I had asked him to; this was about five days before the 911 call. At the time she was "doing OK," he said, but acknowledged her decline in speech. How was *he* doing, I asked. "I'm OK, too," he said, but I remember thinking, 'he doesn't sound OK.' He sounded very depressed. Did he understand what was going to happen one day, perhaps soon? Yes, he said. He could see it coming. Did he and Mrs. Miraly make any decision about what we had discussed in the hospital? No, not yet. Does she want to be kept alive on a machine if her breathing because severely impaired? Almost inaudibly, he said "no, I don't think she'd want that."

But she made no living will and I never did get to talk to her.

* * *

I pull the car up to Doctor's Parking behind the ED and rush in. It is about 1:00 a.m. and the place is quiet, maybe four or five patients are being seen. I ask the triage nurse Rebecca where my patient is.

"Room five. Dr. Stanton's in there now." I can see the waiting area where Mr. Miraly is sitting but he can't see me. I go straight to room 5.

She is lying on a hospital stretcher, her head turned to the left. An ED nurse is taking her vital signs and Dr. Stanton is listening to her lungs. She is pale, gaunt, ashen, diaphoretic, with sunken cheeks and breathing fast: all the ways to describe a patient acutely and chronically ill, perhaps even dying. She is at least 15 pounds lighter than last month.

I introduce myself to Dr. Stanton and we shake hands. We have seen each other in the hospital but not formally met. He is in his early 30's, and I recall that he only recently finished the training program in Emergency Medicine. This makes him an attending physician, and as responsible for his area, the ED, as I am for mine, the ICU.

"Larry, we've just started some IV fluids," he says. "We've sent a blood gas and she just had a chest x-ray. She's only been here a few minutes. I think she's going to need intubation. What's the situation? She's only fifty-seven?"

He means: She looks a lot older than 57, maybe 77. Do we intubate this young/old patient with known ALS, connect her to a breathing machine, and send her to intensive care? Or do we not intubate, and send her to a regular medical ward where she will be made comfortable but surely die soon? Had I not called ahead she would have already been intubated, no doubt, but he has held off waiting for me. He feels responsible for her care at the moment, and wants to get the issue resolved. He doesn't want her to die in his ED. Above all he wants to do the right thing.

ED doctors are as sensitive to end-of-life issues as any group around. They deal with them frequently, and often under difficult circumstances such as the one now presented by Mrs. Miraly. The problem is that the vast majority of chronically ill, dying patients who 'hit the ED' come by ambulance, without any advance directives, or even any accompanying relatives. It is also unusual for any doctor who knows the patient to call the ED in advance, much less show up in time to be of any help. Most of the time the patient is simply brought to the ED by an ambulance that was called by relatives or nursing home personnel. Even if the patient has executed an advance directive at some point, the document almost never accompanies the patient; it is usually locked up somewhere, with other valuable papers.

So when patients look like they're about to die, the ED doctors intubate. They are trained to save lives, and sometimes decisions have to be made before the proper documents or knowledgeable relatives can be located. This is why, frequently, nursing home patients dying a natural death end up

getting intubated, even if the patient is 90 years old and riddled with cancer.

The ED is a way station and patients stay there only briefly. Invariably the resuscitated terminal patient is sent to the ICU, where a different set of doctors has to sort everything out. What's the diagnosis? Has she established any advance directive? Can he speak for himself? (Usually not, by this point). Is the family available, and what are their wishes, expectations? And, by no means unimportant, what is our assessment of the prognosis? Do we think the patient is *truly* terminal?

The ethical ideal of an "autonomous patient" making cogent end-of-life decisions is rare. Most patients – most people before they are patients in any sense of the word – do not make end-of-life decisions, set up advance directives, or even discuss the issue with loved ones. Why should they? People have been dying for millennia without making "end-of-life" decisions. Why all the emphasis now?

The answer is twofold. First, because American medicine has the machines and the staff to manage them. Second, because circumstances and the aforementioned culture make it impossible to always use such vast resources wisely. In the last two decades, in this country at least, an infrastructure has developed to keep dying people alive at all costs. Not just the acutely ill patient who will eventually recover, but the terminally ill who will not recover, who will die without ever getting "off" the machines. Machines to breathe the patient, dialyze the blood when the kidneys fail, and feed the stomach, machines that can extend the dying process for a long, long time. And not just machines, of course, but people to service them *and* care for the patients (most of whom remain in the hospital or end up in a nursing home).

Because this infrastructure is widely available, and mostly paid for by third parties, it often ends up being used when it should not be. "Should not be," of course, is a value judgment. Let's just say the machines are often used when neither the patient nor the family wishes it, and when the machines will not return the patient to any functional state.

Advance directives are supposed to forestall this inappropriate technological juggernaut, but they are largely ineffective. Too few people execute them, and even then the wording is simply too general to be of much help.

* * *

"Yes, she's fifty-seven, Roger. And I honestly don't know the situation. I've talked to them frequently about this happening from ALS, told them what to expect, but they haven't made any decision yet."

"How are they leaning? Because I think we have to decide one way or the other, and soon."

"I know. Let me talk to Mr. Miraly again. I'll take responsibility for

this."

"OK, Larry. Let me know what's what."

Roger leaves to see someone else in the ED, and I move to the bedside to examine Mrs. Miraly more closely. I listen to her heart and lungs, check her abdomen, review the chest x-ray. Everything points to pneumonia in one lung, presumably from aspiration; her muscles are too weak to allow her to cough and clear secretions, and these secretions have become infected. It is a common scenario.

Rebecca comes in with her blood gas results. They confirm respiratory failure – her CO_2 level is very high – and the need for intubation and a ventilator if she is to have a chance of survival.

I look at my patient and rhetorically ask, "Mrs. Miraly, do you recognize me?" She stares past me, unable even to make eye contact.

"Can you hear me?"

She does not respond. Covering her face is a loose fitting, lucent green oxygen mask. The mask hisses quietly. The only other sounds are her own breathing, and the beep-beep-beep of her oxygen monitor. Her cognition is gone and she can no longer participate in end-of-life decisions. I chance that, in the next few minutes at least, she will not stop breathing, and I go out to meet with Mr. Miraly. (In the meantime Mrs. Miraly's nurse will check vital signs.) This time I would rather talk to Mr. Miraly away from her bedside.

* * *

"Mr. Miraly, Mrs. Miraly looks very bad. She is dying. If we intubate her – put that tube in her throat like she had last month – she will end up on the breathing machine the rest of her life. You know we talked about this possibility. Did she make any decision about this? Did you two ever discuss it?"

There is no answer. His face is weary, eyes slightly bloodshot. He has a large bushy mustache and a full head of black hair, which make him look younger than his wife, even before she became ill. I have time to study his face since he only stares at me after my question, as if I had another one to follow.

I don't. After perhaps half a minute he responds with a non sequitur in that low, almost inaudible voice. "She was a beautiful woman, Doctor Martin. A beautiful woman. You didn't know Angela. Where did this come from, this Lou Gehrig disease? What did she do wrong? She doesn't drink, she doesn't smoke. Why should she live connected to any machine? But how can I let her die like this?"

"It may be best to go this way, Mr. Miraly. Most patients with this disease die of pneumonia, and perhaps that is the best way. We can give her antibiotics and make her comfortable, and sedate her so she doesn't feel

short of breath, so she won't feel any pain."

He shows me a picture taken 10 years ago, of the two of them at a wedding. In the photo she is thin and pretty and they are both smiling, happy. The thought enters my head that I don't know these people at all, only her disease, and how it is destroying her body.

"Did you talk to your children about this, I mean about making this type of decision?"

"They don't know she's here now. I don't want to call them now. It's one in the morning." He has not answered my question, so I guess the answer is no.

"Mr. Miraly, should we make her DNR [do not resuscitate], and just treat her with antibiotics and no intubation? Is that what she would want us to do, Mr. Miraly? Or does she want to live on the breathing machine?"

"I don't know. What do you think we should do?"

"Mr. Miraly, we've talked about this before. It's not my decision. I think you have to understand the options. She'll die if we don't put the tube in, but if we do and she recovers from this pneumonia, she will remain on the breathing machine. Then she'll be bedridden, but alert. We'll have to do a tracheostomy to put the breathing tube in her neck, and place a feeding tube into her stomach. She'll need a nursing home. And she'll die eventually, with all these tubes. You need to realize this is how it will be. Is this what she would have wanted? You said she didn't want to live like that?"

"I don't know," he mumbles.

"Did you talk about it with her, at all?"

"No, I don't think so."

I remain silent. After another long pause he says, "Doctor, these last few weeks have not been good to her. I don't have my Angela anymore. Do whatever you think is best."

I suddenly appreciate that he is no longer involved, that I am getting nowhere. Somewhat surprisingly, he has not asked to see her in the treatment room. He knows she is yet alive, and I assume he is fearful of what he'll see; he can no longer deal with what has happened to them. For different reasons, patient *and* husband are each incapable of making end-of-life decisions. If words alone are important, I can obtain any answer I wish to hear. By posing questions a certain way I can get him to mouth words, but they will be without conviction. Realistically, there is no reason to continue this dialog. I am at the point where IT IS UP TO ME. Let her die of natural causes, humanely, or subject her to machines, tubes, life support for as long as her body can hold out.

"Then I'll decide, is that what you want Mr. Miraly? Do you want for me to decide?"

"Yes, you decide, doctor. Do what's best, doctor, please."

The waiting room is virtually empty, and we are sitting side by side on a

long row of hard plastic seats. I need a witness and call Rebecca over; for the moment she is not busy. Apart from registering Mrs. Miraly upon arrival, Rebecca does not know her problem in any detail (a different ED nurse is providing care as I speak with Mr. Miraly). Rebecca sits down on another seat directly opposite us. Like Roger she is also young, in her late 20's. She has worked as triage nurse in the ED for about a year. I don't know if she has ever been involved in end-of-life discussions (I doubt it, at least while a triage nurse), but she is a professional and I just need someone to listen in. I briefly explain Mrs. Miraly's condition, and our conversation, then resume my questioning.

"Mr. Miraly, I want this nurse to hear you, because this is very important. You understand your wife is dying of a terminal condition?"

He looks at me and nods yes.

"She has never stated her wishes, of what to do when the end comes, is that right?"

He looks at Rebecca, then me, and again nods yes.

"Because she has a terminal condition, if I think it's best to just keep her comfortable, and not to place her on life support machines, not to put a tube into her throat, then you go along with that Mr. Miraly?"

He forsakes the nod and says, "Yes, if that's for the best."

"So it's OK, then, Mr. Miraly, *not* to put Mrs. Miraly on life support machines? Not to intubate her throat and connect her to a ventilator?"

"Yes, if that's what's best for her." Did he emphasize the "if"? I seek to clarify further.

"You understand that we'll keep her comfortable, make sure she doesn't suffer, and that she'll still be cared for in the hospital until the very end?"

"Yes, if that's what you think is best," he whispers. There is no emphasis on "if," no emphasis on "best," no emphasis on anything. He is staring at her photo, and is no longer looking at me or the nurse. I ask Rebecca if she heard everything, and if she has any questions for me or Mr. Miraly. This is not a deposition, only a conversation that I want witnessed. Rebecca affirms she heard him clearly and that she has no questions. I thank her and she returns to the triage desk.

The ED is quiet. In the distance I hear an ambulance siren, and closer in the muffled voice of the hospital operator paging a physician. I am thankful that we are alone; no one is nearby, no one is observing us. I grab his free hand and hold it tight with both my hands. His hand is limp. His head is bowed and he is crying. I am about to cry, too, but hold it back.

I envision us as posing in some modern tableau – two vulnerable people, patient's husband and doctor, one crying and one trying not to. If the scene was painted, perhaps by a post-modern Normal Rockwell, it would be titled "Decision Made In The ED."

We sit still for a full minute. No more words pass between us. His mind has retreated to some happier time long past. I am not so lucky. My

mind is rooted in the present, the god-awful decision-time present. I search for precedents and realize there are none, at least none just like this. The disease may have an ineluctable natural course, but every case is different, with singular nuances of family, doctor, hospital, and location in space and time. I think to myself: 'We are all unique upon the earth; disease does not change this.'

I let go of his hand. He remains silent. I get up and walk toward Mrs. Miraly's room. On the way I glance back at Mr. Miraly. He is not watching me, only staring at his wife's picture.

My patient remains obtunded, her breathing fast and labored. At the moment, no one else is in the room. Fluid is dripping into her veins. I glance at the vital sign sheet. Her blood pressure and heart rate are holding up. I call Dr. Stanton and Mrs. Miraly's nurse into the room, and explain my conversation with Mr. Miraly, and how he has agreed to do whatever I think is right for his wife. I explain how difficult this has been for both of us, and what my decision is. They nod approval.

And then I intubate her.

-- END –

23. Shock Him!

John Capowski, 51, came to the hospital for an altogether different problem than what brought me to his bedside. He showed up in our emergency room weak and lethargic, with a three day history of nausea, vomiting and diarrhea. He came because he couldn't keep anything down, and also because it was the middle of the night and Mrs. Capowski was afraid. She insisted he see a doctor and drove him to our ER. They arrived about 1 a.m.

The previous day he had missed work, calling in sick to the car plant where he worked as assembly line foreman; the line makes engine blocks for several car models. His job was not difficult, he liked the work and was seldom absent. A second day absent, though, and he would have seen a company doctor, if for no other reason than a return-to-work note. He rarely saw his own physician, certainly not for anything routine. And he wouldn't think of calling his doctor at midnight, so the ER was a convenient choice.

To the ER physician on duty Mr. Capowski did not seem acutely ill. Vital signs were normal, although his blood pressure was slightly on the low

side. But vital signs like blood pressure and heart rate are not the whole picture. Despite his size – a muscular 5'11", 210 lb – he appeared wan and listless, sick enough to warrant further investigation. Vomiting and diarrhea can deplete the body of electrolytes and the only way to know for sure is to measure the blood levels. (And who would want to tell Mrs. Capowski to take him home? She hovered over him with an air of insistence: 'do something').

Sure enough, Mr. Capowski had a low blood sodium and potassium, meaning he had lost a significant amount of electrolytes. He needed hospitalization, a recommendation as much to his chagrin as to his wife's relief. Other tests in the ER, including chest x-ray and EKG, were normal, so he was admitted with the diagnosis "Probable viral syndrome, dehydration, hyponatremia [low sodium], hypokalemia [low potassium]." The plan on admission was "fluid replacement and monitoring of electrolytes." The ER doctor started an intravenous solution of sodium and potassium and sent him to the ward.

Mr. Capowski reached Tower 9 about 5 a.m. Four hours in the ER is not a particularly long delay, especially since treatment was started there. By the time of transfer he was feeling much better. The night shift nurse on Tower 9 noted he was "stable . . . not short of breath" and "has no complaints."

From then until 8 a.m. things went well. Mr. Capowski slept a little, made one call to the nurse for an extra pillow, and otherwise created no stir. Shortly after 8 a.m. he got up to go to the bathroom and halfway there slumped to the floor. A nurse in the hallway, Nancy Whitehead, heard a crashing sound (probably the IV pole hitting the floor along with the patient) and went in to check. She found Mr. Capowski on the floor, unresponsive, face down. She tried to arouse him. Nothing. She instinctively felt for his carotid pulse. Nothing.

One second he was standing, breathing, living; the next second, no heart beat, no respirations, a lifeless hunk on the floor.

* * *

People die suddenly and non-traumatically all the time – several hundred thousand a year in this country alone. It usually comes from cardiac arrest; the heart suddenly stops beating, followed immediately by cessation of breathing. Sometimes a rescue squad arrives in time to revive the victim, but far more often there is nothing to revive when help does come; the heart is irreversibly gone and Mr. Jones or Ms. Smith is "dead on arrival" to the closest hospital. Sudden death from cardiac arrest happens to people at work, on the golf course, in airplanes, at shopping centers, at home. It can happen while exercising, sitting or sleeping. Usually the victims are older and have some history of heart disease, but many are younger (less than 50)

and have no known heart problem.

Basic Cardiopulmonary Resuscitation (CPR) has been a staple of Red Cross teaching for decades. It is taught in schools, churches, businesses, literally any place people will gather for a two hour session. The lessons are simple. Come upon a victim? Establish unresponsiveness by shaking him (or her). Without this crucial first step, you may give CPR to someone merely napping. Unresponsive? Immediately call 911 or tell someone to do so. Then open the victim's mouth and give two slow mouth-to-mouth breaths. Next, feel for the carotid pulse in the neck region, just behind the angle of the jaw. No pulse? Pump on the lower breast plate (sternum) 15 times with the heels of your hands. Then give two more breaths, followed by 15 more pumps. Repeat the sequence (two breaths, 15 pumps) until there is a pulse and breathing, or you become exhausted, or help arrives.

There are some dramatic saves with basic CPR but the technique is, by itself, not sufficient for the true cardiac arrest victim. Chest pumping and mouth breathing provide a fraction of normal heart and lung function, and can only be sustained for minutes. So CPR's main value is in buying time until people can come with advanced life support equipment. But if the rescue squad doesn't arrive quickly the situation is hopeless. Or worse. The heart rhythm may return too late, leaving the victim alive but severely brain damaged, a human vegetable relegated to institutional care. It happens.

No doubt in some cases the heart may be ready to quit, and a resuscitation attempt is futile. The organ may be so diseased that any measure, short of instant heart transplant, is bound to fail. But for most sudden death victims there is probably much life left in the ticker *if only it could be re-started before the brain is damaged*.

A dead car battery means a dead car, but if the battery only needs to be jump- started the car might be able to go another 100,000 miles. Without energizing the battery no amount of gas or engine repair will give life to the car. Of course car batteries can be dead a long time before coming to life again; in cardiac arrest the victim has, on the outside, only 4-6 minutes. After that, brain damage from lack of oxygen is irreversible (with rare exceptions, such as after near-drowning in cold water).

There is another difference between the heart in cardiac arrest and a dead car battery. To understand what happens in most sudden death cases, accept the fact that the heart doesn't just shut down; it begins to violently *wiggle* for a few minutes, *then* shuts down. Only during those precious few minutes *before* total standstill does the victim have a chance.

The heart is an amazing bundle of muscle and nerve fibers. These fibers, distributed in discrete bundles throughout the organ, have automaticity, which means they fire off automatically until the end of life. You can't move an arm unless your brain sends a signal to the arm nerves to move the arm muscles, but your heart muscle will contract whether you think about it or not. Nerves bundles in the heart are not controlled by the

brain.

Each firing of the cardiac nerves causes one contraction or heart beat, and with it the heart muscle pumps blood out the aorta, to the rest of the body. The adult human heart beats about 70-80 times a minute for the life of its owner; this comes to 37-42 million heart beats a year, or *billions* of beats over the average life span.

Most cases of cardiac arrest occur because the heart's nervous system suddenly runs amok. The cardiac rhythm changes from regular to a *chaotic, purposeless wiggling* called *ventricular fibrillation*. In anyone not ready to die, not expected to die, ventricular fibrillation is the most feared diagnosis imaginable. From the onset of VF the victim becomes instantly unconscious; in only a few minutes the heart in VF will stop all activity and be forever motionless. A totally motionless heart is called asystole, literally absence of systole or contraction of the heart. The fibrillating heart can be resuscitated; the heart that fibrillates and then goes into asystole cannot (with rare exception).

The chaotic nerve firing of ventricular fibrillation emanates from the ventricles, the large pumping chambers of the heart. VF is not to be confused with the relatively benign rhythm *atrial* fibrillation, a common malady in older people. In AF the smaller chambers (atria) fibrillate but the ventricles beat normally. People can live for years with AF; no one can live more than a few minutes with VF.

The heart's visual appearance in ventricular fibrillation has been likened to a bag of worms. In VF every heart muscle fiber beats to its own rhythm without regard to the one adjacent; there is no coordination, no synergy, no *pumping action*. As a result, no blood leaves the heart during ventricular fibrillation and there is no pulse. Sudden death is not always from VF but VF always means sudden death – unless it is quickly, quickly reversed.

Why does a heart suddenly go into VF? Heart attack is probably the most common reason, a sudden lack of blood supply to a critical part of the heart tissue. People who die of "heart attack" usually have VF first, then die.

But lack of blood supply can also manifest as a non-fatal *arrhythmia* or abnormal beating of the heart. Not surprisingly, many cardiac arrest victims have a history of arrhythmia in the preceding months or years. (Fortunately the converse is not true; most cardiac arrhythmias, which are exceedingly common, do not presage VF.)

Here's what a heart with normal cardiac rhythm sounds like; each thump represents one heart beat.

"**thump--------thump--------thump--------thump--------thump--------thump--------thump--------thump**"

A too-fast heart beat is called tachycardia. It can be regular or irregular. Here's the sound of a regular tachycardia.

"thump-thump-thump-thump-thump-thump-thump-thump-thump-

thump-thump-thump-thump"

Here's a tachycardia where every third beat is abnormal (called trigeminy). **"thump-thump-THUMP-thump-thump-THUMP-thump-thump-THUMP-thump-thump-THUMP-thump-thump-THUMP"**

And a tachycardia where the abnormal heart beats are different from each other *and* occur at irregular intervals:
"**thump-thump-THUMP-thump-THUMP----thump-thump-thump---thump-*THUMP*---THUMP-thump-thump-thump-THUMP**"

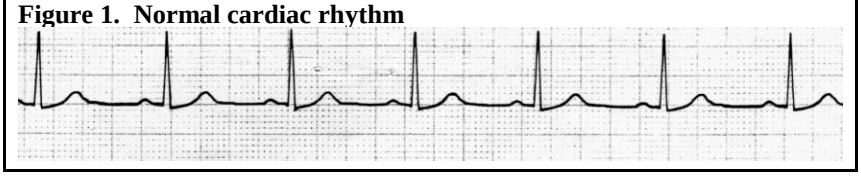
The variations in tachycardia and it's opposite, bradycardia (slow heart beat), are numerous. However, as long as there is a "thump" some blood will be pushed out of the ventricles to the rest of the body. Treatment of arrhythmias depends on the nature of the thumps (diagnosed by the EKG and physical exam) and how the patient feels. Treatment is usually required if the patient experiences discomfort ("palpitations") or the abnormal thumps pump too little blood to the body. In any case doctors have one basic rule: treat the patient, not just the arrhythmia. Sometimes cardiac arrhythmias are best left alone.

VF is never best left alone, unless death is expected. There is no "thump" in the VF heart, no pulse, because the heart wiggles without purpose. If reversal doesn't come within minutes the patient always dies.

The only way to recognize VF is with an EKG machine or other type of cardiac rhythm monitor. These machines, which connect to the patient via wires, sense the heart's electrical activity and provide a continuous tracing of the cardiac rhythm. Given such a device, diagnosis of VF is not usually difficult. In fact the biggest obstacle to recognition is probably artifact, from electrical noise. This noise can be generated by the environment or by wires improperly attached to the patient. It can sometimes look like VF. (When the victim is without pulse or respirations, the rescuer focuses on checking for VF and making sure the wires are properly attached.)

Figure 1 shows a normal heart rhythm taken from an EKG tracing; each narrow, symmetrical spike or deflection is preceded and followed by a shorter, rounded deflection; taken together the three deflections represent the electrical activity of *one* normal heart beat (one "thump"). The distance between the tall spikes in this tracing indicates a heart rate of about 70/minute. (The closer the spikes are to each other the faster the heart beat.) Note in this tracing how every heart beat looks the same.

Figure 1. Normal cardiac rhythm



The cardiac rhythms shown in Figures 2-5 are all abnormal. Each rhythm represents one of the following diagnoses (not in order):

- regular tachycardia
- regular tachycardia
- asystole
- asystole
- rhythm where every second beat is abnormal (bigeminy)
- rhythm where every second beat is abnormal (bigeminy)
- ventricular fibrillation (Mr. Capowski's rhythm).
- ventricular fibrillation (Mr. Capowski's rhythm).

Which is which? Try to answer before proceeding (answers at end).

Figure2.?Diagnosis.

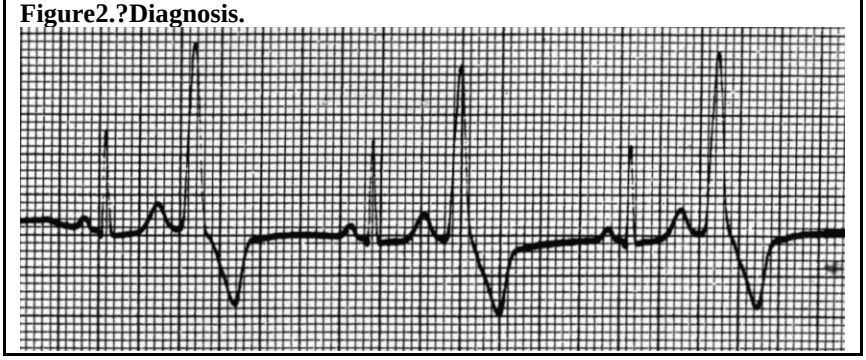


Figure 3. ?Diagnosis.

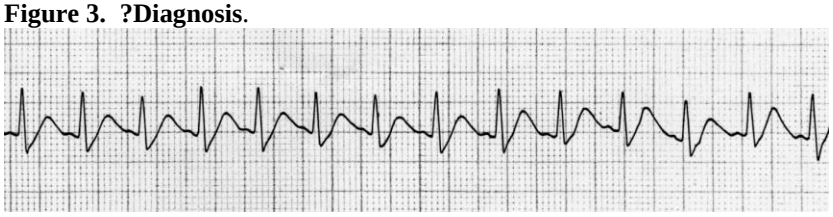


Figure 4. ?Diagnosis

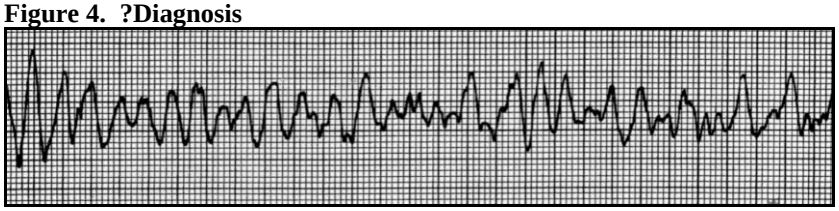
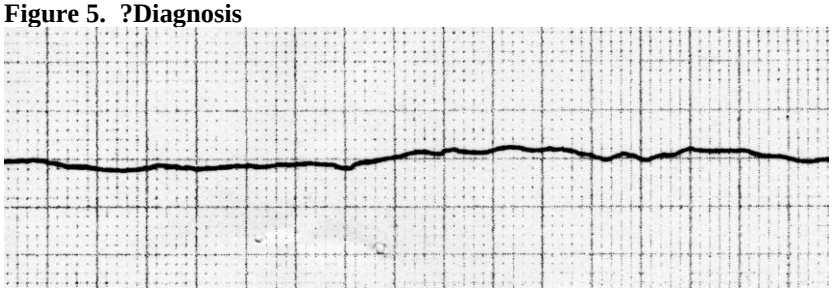


Figure 5. ?Diagnosis



The only therapy for VF is *defibrillation*, literally a jolt of direct current electricity delivered through the chest. This jolt doesn't actually re-start the heart like a car battery; instead it stuns the heart momentarily, stops the VF, and allows the heart's normal nerve firing to resume. Defibrillation requires a machine, a defibrillator, that can be attached to the victim via wires and deliver the shock. The defibrillator also contains a cardiac monitor to display the rhythm. But where do you find a defibrillator on the golf course? In the shopping mall or ball park? In the home?

If the defibrillator was like a fire extinguisher, available in all public places and most homes, probably thousands of people could be resuscitated from certain death every year. But defibrillators are not like fire extinguishers. They are expensive and intimidating to use. Anyone can recognize a fire and use an extinguisher. Hardly anyone without special training can recognize VF and use a defibrillator.

What if defibrillation was part of basic CPR? In fact, it is becoming so, albeit slowly. Several defibrillators are manufactured for use by people trained only in basic CPR (people just like you, reading this story). The devices are called Automated External Defibrillators. AEDs differ from defibrillators used in hospitals (called manual defibrillators) in one important aspect. AEDs analyze the rhythm for you.

With a manual defibrillator the rescuer must do everything: attach wires to the patient, turn on the machine, analyze the rhythm and, if VF, apply paddles to the patient's chest, dial the amount of electric shock and push buttons to deliver it. The process doesn't take a long time, but the real skill is in analyzing the rhythm. If the rhythm isn't VF a defibrillator shock *can*

cause fatal arrhythmia.

With an AED the process is greatly simplified because the machine analyzes the rhythm, and very well. The best AED analysis programs are so sophisticated that they can separate most artifact from VF, and do as well as physicians in diagnosing the fatal rhythm. The rescuer still has to identify the victim, establish unresponsiveness and pulselessness (basic CPR), find the machine, turn it on and attach two large adhesive pads to the chest. (These pads stick to the skin and replace the hand-held paddles that come with manual defibrillators.)

Fully automated AEDs will analyze the rhythm and automatically shock the patient. *Semi-automated* AEDs will analyze the rhythm, announce the finding of VF through audio or a screen display, and advise the rescuer to press a button to deliver the shock. In either case the rescuer doesn't have to hold paddles to the chest; with an AED defibrillation is a hands off procedure. The shock can be repeated as long as the machine reads the rhythm as VF.

The fact that there are several AED models doesn't detract from their ease of use. AED-training is now incorporated into many basic CPR courses, but clearly not enough. Most people don't know about AEDs, most CPR courses don't teach their use, and most public places don't have them available.

Why not? Perhaps society is too accepting of sudden, non-traumatic death. We should think of it as treatable, and by almost anyone. After all, what do you have to lose? Without defibrillation most victims of cardiac arrest stay that way.

What if the maître d' Raphael, in Thomas Wolfe's novel *The Bonfire of the Vanities*, had mobilized basic CPR when his obese customer slumped over from a heart attack? The thought of resuscitation never crossed his or *anyone else's* mind in the crowded restaurant. The diners stay put because, after all, "the orders were in, and the food had begun to arrive..." Raphael's reaction is worse: sheer annoyance at the newly deceased patron. Arriving any minute to his tony establishment is the wife of a foreign dignitary, and this unfortunate incident must not disturb preparations. "The waiters were now skipping over the corpse mindlessly, as if they did it every night, as if every night there were one corpse or another lying in that spot. . ."

Wolfe intended the scene as satire, but it's telling nonetheless. If life imitates art, then an indifferent attitude toward sudden death has become part of our culture. Societal indifference is more subtle than the maître d's, to be sure, but it can be just as lethal. Indifference may manifest as a company policy not to train employees in CPR, or as a government policy not to budget for AEDs in public facilities, or as a citizenry's laissez faire attitude about learning CPR. Yet two facts remain. Thousands of real, not-yet-ready-to-die people slump over dead every year. Many of them could be fully resuscitated.

* * *

Nancy ran into the hallway and yelled: "Call a chest team!" Chest team is our code for cardiac arrest. Different hospitals use different codes, such as "Dr. Heart!", "Code Blue!", and "Code 911!"

The ward secretary punched a special 3-digit number into her phone and the hospital operator instantly answered, "Where?"

"Tower nine."

Elapsed time since Mr. Capowski's fall: 30 seconds. Nancy grabbed the red crash cart standing in the hallway as the loudspeaker boomed:

"CHEST TEAM TOWER NINE. CHEST TEAM TOWER NINE. CHEST TEAM TOWER NINE. CHEST TEAM TOWER NINE."

I heard it. Everyone did, because chest team is broadcast hospital-wide as a signal for specific people to come, wherever they are: the anesthesiologist to intubate the patient, the senior medical resident to help manage the arrest, the respiratory therapist to set up the ventilator, and all doctors in the immediate vicinity. This time I am in the vicinity, three rooms away seeing another patient. I go into the hallway and immediately see all the tumult.

As I enter Mr. Capowski's room there are already two nurses and two medical residents, initiating resuscitation maneuvers. Mr. Capowski has been turned over and is now supine on the hard linoleum floor. Elapsed time from fall: 45 seconds. Nancy is pumping on Mr. Capowksi's chest while one of the residents is pushing air into his mouth and nose with a bag-mask-valve contraption called an AMBU bag. (These devices are safer and more effective than mouth to mouth breathing, and so are universally used by professional rescuers.) Another nurse is hooking up monitor leads from the defibrillator, which now sits on the floor next to the victim. The other resident is feeling for a pulse.

"What happened?" I ask. A dumb question, as everyone is in the midst of resuscitation. The man arrested, that's what happened.

"I just came on," says Nancy, while pumping. "This is a Mr. Capowski. He was admitted a few hours ago with dehydration. I was told he was stable. He just collapsed."

"Do we have any history?"

The night intern who admitted him to the ward is apparently not here. Probably signed off and taking a shower. Another resident, who entered right after me, begins reading through the medical chart out loud. "He came in just this morning. Let's see, no history of heart disease. On no heart or other meds. In fact, pretty much no medical history. He works in a factory. Smokes a pack a day, for 30 years. Drinks alcohol socially. His sodium's low, 129, K [potassium] is only 3.3. They gave him some K in the ER. His EKG was read as unremarkable down there."

"Let me see it." His EKG is in the chart and it looks normal. I begin to grasp the picture. Mr. Capowski wasn't expected to die. This is not terminal cancer or end-stage heart disease.

"He's in V fib!" yells the nurse who attached the chest leads. She instinctively reaches for the defibrillator paddles to apply to his chest.

Time from fall: one minute 40 seconds. We are on a roll. Most in-hospital chest teams take several minutes for someone to bring the defibrillator to the bedside *and* attach the leads *and* read the monitor rhythm. Time is life and, so far, this is good time.

Meanwhile Nancy is still pumping and the resident is still AMBU bagging. More people enter the room, too many in fact. "Chest team" always brings in too many people. Better too many than too few, but sometimes crowding is a problem. Everyone who enters has a question, or a suggestion of some sort (where's the oxygen? does he have an IV? how long's he been down?). Nancy, sensing impending chaos, quickly turns her head to me and without hesitation intones, "Dr. Martin, you're in charge."

Me in charge? I just walked in, and for a minute have sort of just stood by while others do the work. Why am I in charge? Of course, resuscitation by a bunch of people requires that someone be in charge; otherwise conflicting orders tend to be yelled by many lieutenants. And, someone has to be ready to make the most important decision of all: when to stop.

Usually in charge is the senior person trained in Advanced Cardiac Life Support. (ACLS incorporates defibrillation and use of cardiac medications.) Trained, of course, means simply having taken the American Heart Association's two-day ACLS course and knowing what to do. Any of the medical residents could be in charge, since I am no more knowledgeable in ACLS than they are, but to quibble at this point is self-defeating. Nancy is going by the book and wants to establish a leader, and I understand. My thoughts on this matter take about three seconds.

I notice the paddles are now applied to his chest.

"OK, give him 200 joules," I say. Joules are the energy units for electric countershock. In VF we start with 200 joules, and if that doesn't work go to 300 joules and if that doesn't work, 360 joules.

"Stand back! Hands off!" yells the nurse holding the paddles.

Rescuers have to keep hands off the patient and all equipment touching the patient, or they can be shocked by the defibrillation energy. So CPR always stops when the shock is delivered – at that moment there is no pumping on the chest, no bagging. Hands touch the shocking paddles only: one held against the upper right chest beneath the clavicle and the other pressed under the left nipple, over the heart. Buttons on both paddle handles are pressed simultaneously and the jolt of electricity is delivered to and through the victim's heart.

The buttons are pressed, the jolt delivered. Zap! Mr. Capowski's body jerks. All eyes turn to the defibrillator monitor. Nothing seems to have

changed. The monitor shows he's still in VF. Time elapsed: two minutes 20 seconds.

The anesthesiologist walks in, along with the respiratory therapist. There are now 10-12 people in the room. Only five or six are needed, tops; the anesthesiologist is one of them. Observing that the shock has done nothing, she asks if we want her to intubate the patient. Placing an endotracheal tube in his throat would certainly allow for better ventilation and oxygenation then using a face mask.

Intubate now or shock again? The decision is easy. There is a universal algorithm for treating VF, taught as part of ACLS. The thinking about VF goes like this. Only electrical defibrillation will correct the problem; nothing else seems to work. Two hundred joules frequently does the job, but sometimes only a higher dose will work when the lower dose fails. Also, repeat shocks seem to work better even when the dose is kept the same. Any delay in providing shocks can be fatal. Therefore, don't waste time trying to intubate (even in the best of hands it could take a minute) in lieu of giving initial shocks. *First thing to do is give three successive shocks: 200-300-360 joules.* Do this before any drugs are given, before an attempt at intubation, even before placing an intravenous line.

"No, he needs another shock," I respond. "Go to 300 joules." The defibrillator knob is turned a notch to 300.

"Everyone off." The paddles are applied against his chest.

Zap! His body jerks again.

"Let's check the rhythm." Still VF. Three minutes 10 seconds have elapsed. Nancy bends down to resume chest pumping but I tell her to hold off.

"Give him 360." This is the maximum. If he doesn't respond, then we'll resume CPR, go to intubation, IV epinephrine and another shock at 360. Then more epinephrine and another 360. On and on. The longer he goes without responding the less likely he'll respond.

Zap!

One-two-three seconds. The VF rhythm disappears and we have. . . a flat line! Asystole! But before I can yell for something else to try the flat line is interrupted by regular blips.

"We have a rhythm!"

Again, all eyes are on the monitor. In the space of 10 seconds the chaotic wiggling of VF has changed to asystole and then to a regular heart beat. Not a normal rhythm to be sure, but one that *gives a pulse*. We can feel it in his groin. He is still unconscious but he has a pulse and a blood pressure. Now I let the anesthesiologist intubate him.

* * *

An hour later he is in the intensive care unit, under my care. And he is

waking up! Mrs. Capowski is at his bedside. (A nurse called during the chest team and told her he "suddenly collapsed" and "the doctors are working on him." She arrived about a half hour later, not knowing if her next step would be to bury her husband.)

We have much work to do on our new patient but she needs to be with him, now. Who knows what the next 24 hours will bring? His heart could stop again and he could die next time. We are all a bit uneasy. I have asked a cardiologist to see him right away and help us determine where to go next; I assume he will need cardiac catheterization as soon as possible. Considering he was unconscious for less than four minutes, and his heart was manually compressed during some of that period, Mr. Capowski has an excellent chance of being mentally fine.

I am smug, for this was truly a miraculous save. A combination of luck and nursing skill. In fact it is one of the very few times I have been witness to a successful in-hospital cardiac arrest outside the intensive care unit or emergency department. In-hospital resuscitation is usually dismal for several reasons. First, people who develop VF on a hospital ward are typically elderly and/or debilitated and/or with a terminal disease.

Successful chest teams in the setting of advanced disease are rarely happy events, even when the patient survives. Hospitals routinely resuscitate chronically ill, debilitated patients but save very few; most die later, during the same hospital stay. Survivors who leave the hospital are usually relegated to long term nursing home care, often connected to a breathing machine.

Another reason for dismal outcome has to do with patient location. In the ICU patients are monitored continuously whereas on the regular medical ward, only every few hours or so. Thus a patient can go into cardiac arrest *without anyone knowing*. It is not at all unusual for a nurse or orderly to come upon a patient pulseless and cold.

A third reason is response time. Even when the arrest is promptly identified it may take several minutes to get the crash cart and attach the defibrillator wires. In non-patient care areas, such as the waiting areas, parking garage, hospital lobby – all places patients have arrested – resuscitation can take much longer. (Some hospitals are placing AEDs in these remote areas, to be used by anyone properly trained.)

Mr. Capowski's luck was in falling where he did and someone hearing him fall, immediately, someone who was trained in what to do and able to do it without hesitation. The chest team worked like it's supposed to.

* * *

In the ICU Mr. Capowski's post-arrest EKG showed evidence for a myocardial infarction, a heart attack, no doubt sustained while walking to the bathroom. Most likely his smoking and the low electrolytes, plus some type

of underlying heart disease, all contributed to VF. Because of VF he did require emergency cardiac catheterization, carried out the same afternoon. The cardiologists found a critical narrowing in one coronary artery, and they were able to open it up with balloon angioplasty. During the procedure he received intravenous xylocaine to prevent recurrence of VF (the drug was started immediately after resuscitation). Also, a heart surgeon was available in case angioplasty failed and coronary bypass became necessary. Fortunately, it didn't.

Mr. Capowski left the hospital just four days later, on his own feet. By then at least six different doctors and nurses had regaled him about the event, including me. I emphasized prevention. ("You know your heart stopped, it could happen again, you really have to quit smoking.") Others commented in awe, hinting at his miraculous recovery. ("Do you remember anything that happened? Did you feel anything?") And others were simply clinical and direct. ("Your heart stopped due to a coronary blockage; the artery is open now but you must stay on this [heart drug] indefinitely. We'll see you in the office in three weeks.")

Best selling books have been written about near death happenings, the experience often framed in religious and mystical overtones. But to Mr. Capowski, he might as well have been merely sleeping; he remembers nothing from the time he got out of bed on Tower 9 until he woke up in the ICU. He is not particularly religious, and shrugs off all the attention with a 'That's-my-15-minutes-of-fame' kind of attitude. He had glimpse of neither heaven nor hell and admits to no "experience." As for any lasting effects, Mrs. Capowski swears her husband has the same low key, taciturn personality he had before hospital admission.

Was Mr. Capowski resuscitated from death or near death? During resuscitation was he sleeping or unconscious or dead? Questions like these can make you glassy-eyed with metaphysical thoughts. From a pure physiology perspective all that happened is his heart stopped pumping. During this period his brain had sufficient oxygen (due to basic CPR) so that he suffered no loss of cerebral function.

The day of discharge I went in to say good by. By then "your heart stopped" was almost a cliche, and I didn't bring it up. It was clearly on his mind, though. Not because he planned to write a book but because (I suspect) he was plotting out his post-hospital life style.

"My heart really stopped that day, huh doc?"

"Yes, it did."

"I'll be damned. I feel great now."

ANSWERS

Figure 2. bigeminy

Figure 3. regular tachycardia

Figure 4. ventricular fibrillation
Figure 5. asystole

-- END –

GLOSSARY

AIDS - acquired immunodeficiency syndrome; in AIDS the body's infection-fighting immune system is severely depressed and altered. AIDS is caused by the human immunodeficiency virus (HIV).
angiogram - a test whereby dye is injected into a blood vessel to outline the vessel and any abnormality within it. See pulmonary angiogram.
apnea - absence of breathing; an apneic episode or spell is a short interval of not breathing.
arterial - pertaining to the arteries, e.g. arterial blood.
arterial blood gas - refers to the pressure of oxygen and/or carbon dioxide in arterial blood; abbreviated ABG. An 'ABG' test routinely measures pressures of both gases, along with the level of blood acidity.
arterial line - a thin tube inserted into a patient's artery, usually the radial artery, for purposes of monitoring blood pressure or drawing frequent arterial blood gases. This technique is only used in intensive care units or in the operating room.
artery - blood vessel that carries oxygen-rich blood from the heart to the body's organs and tissues.
artificial ventilation - method of supplementing or taking over a patient's breathing with a machine (ventilator). The patient is connected to the ventilator via an endotracheal tube inserted through the mouth or nose.
artificial ventilator - see ventilator.
autoimmune diseases - a large and heterogeneous group of diseases characterized by altered immunity; usually, antibodies form in the blood directed against some part of healthy tissue

biopsy - removal of a piece of tissue from some part of the body for diagnosis.
blood gases - general term for carbon dioxide and oxygen in the blood; see arterial blood gas.
bronchitis - inflammation or infection of the airways (bronchi).
bronchodilator - a drug that relaxes airway smooth muscle and helps open up narrowed airways; useful in treating asthma.
bronchoscope - thin, flexible tube used to perform bronchoscopy; useful to diagnose many pulmonary conditions.
bronchoscopy - procedure whereby a thin, flexible tube (the bronchoscope) is inserted, via the mouth or nose, into the lungs; used to visualize the airways and diagnose many lung diseases. A biopsy can be done through the bronchoscope.
capillary - the smallest blood vessel. Capillaries go to all organs to bring vital oxygen and take up carbon dioxide; in the lungs the process is reversed:

fresh oxygen is taken up and carbon dioxide excreted.
carbon dioxide - colorless, odorless gas, a byproduct of normal metabolism; abbreviated CO_2. Carbon dioxide is excreted by the lungs through the natural process of ventilation.
catheter - a thin, plastic tube that can be inserted into part of the body, such as a blood vessel or the bladder.
catheterization - general term for inserting a tube into a blood vessel. In cardiac catheterization a thin tube (catheter) is inserted through a vein or artery and into the chambers of the heart.
coronary - pertaining to blood vessels that serve the heart muscle; so-called because the coronary vessels encircle the heart like a corona.
coronary care unit - area of hospital for patients with acute heart disease, including suspected or diagnosed heart attack.
CPR - Cardiopulmonary resuscitation, the series of steps done to revive a person who has stopped breathing and/or whose heart has stopped beating
CT scan - Computerized tomography scan (also sometimes called CAT scan for computerized axial tomography); a sophisticated x-ray technique which can "slice" any section of the body to reveal details of anatomy not seen with conventional x-rays.
coumadin - a medication that makes the blood less likely to clot; used to treat many medical conditions, including pulmonary embolism; can only be taken by mouth.

defibrillator - machine that can provide electrical shock to the heart in order to convert the cardiac rhythm to normal**.**
dialysis - process of cleansing the blood of toxins; in *hemo*dialysis, used to treat kidney failure, blood is removed through a vein, passed through a special filter that removes the toxins, then returned to the patient.
diuretic - a drug that promotes urine flow; a diuretic can be taken by mouth as a pill or administered by the intravenous route.
dopamine - an intravenous drug used to raise a patient's low blood pressure. See pressors.
dyspnea - shortness of breath.

emboli, embolism - when a blood clot moves from one part of the body to another; in pulmonary embolism the clot moves from some region of the body to the lungs.
emphysema - a chronic pulmonary disease, usually due to smoking, that leads to shortness of breath and blockage of airflow.
encephalitis - inflammation of the brain; see encephalopathy.
encephalopathy - a general term for confusion due to global brain disease. There are many possible causes including inflammation (encephalitis) and lack of oxygen.
endoscopy - general term for insertion of a flexible, diagnostic tube

(endoscope) into a hollow organ; gastrointestinal endoscopy involves inserting an endoscope into the stomach or intestines.
endotracheal tube - a hollow plastic tube, approximately a foot long and a centimeter in diameter, inserted through the mouth or nose and into the trachea. It is used to facilitate artificial ventilation.
esophagus - hollow tube that connects the mouth with the stomach.
exsanguinate - to bleed out, hemorrhage.

gastroenterology - specialty of medicine involved with diagnosing and treating gastrointestinal (stomach and intestinal) disorders; a specialist in this field is a gastroenterologist.

hematocrit - percentage of blood volume comprised of red blood cells (oxygen-carrying cells); normal range for hematocrit is 38-45% in women, 42-50% in men.
hemodialysis - see dialysis.
hemoptysis - coughing up blood.
heparin - a medication that makes the blood less likely to clot; used to treat many medical conditions, including pulmonary embolism; is given intravenously or subcutaneously.
HIV - human immunodeficiency virus, the virus responsible for causing AIDS.
housestaff - the interns and residents in a teaching hospital; also spelled house staff.
hyperthyroid - elevated level of thyroid hormone, the hormone that regulates metabolism.
hyperventilation - over-ventilation or over-breathing. Hyperventilation is accompanied by a reduced carbon dioxide level in the blood.
hypothyroid - low level of thyroid hormone.
hypoventilation - under-ventilation or under-breathing. Hypoventilation is always accompanied by an elevated carbon dioxide level in the blood.
hypoxemia - low oxygen level in the blood. Hypoxemia can manifest as either a low oxygen pressure (PO_2) or a low oxygen saturation; see PO_2.

immunosuppressed - when the body's immune system for fighting infection is suppressed or altered; this is the principal problem in AIDS patients. Immunosuppression is also found in many other situations, e.g. during treatment with cancer drugs.
insulin - a hormone made by the pancreas, necessary to allow glucose to enter the cells; a lack of insulin leads to diabetes.
insult - in medical terminology, refers to damage or injury to a part of the body, e.g., an insult to the liver
intravenous - refers to route for medication or fluids given directly into a vein.

intubation - the placement of an endotracheal tube into the patient's airway, usually for purposes of providing artificial ventilation; see artificial ventilation.

meningitis - inflammation of the meninges, the thin membrane that covers the brain and spinal cord.
MICU - medical intensive care unit; area of the hospital for acutely ill patients, excluding those with surgical problems or primary cardiac disease.
Munchausen - name given to a patient who fakes illness in order to gain medical attention or admission to the hospital; after Baron von Munchausen, an 18th century teller of tall tales.
mycoplasma - a bacteria-like organism that can cause pneumonia.

O_2 - Chemical symbol for oxygen.
opportunistic infection - an infection by an uncommon organism (may be a bacteria, virus or fungus), one that takes advantage of a patient's suppressed or altered immunity (hence opportunistic), such as is found in AIDS patients.
overdose - general term for taking an excess of medication; an overdose can be intentional (e.g., suicidal), or accidental (e.g., swallowing too many pills for headache relief).
oxygen - essential element of life; a colorless, odorless gas that comprises 21% of earth's atmosphere. Abbreviated O_2.

Pickwickian syndrome - term used to characterize a patient who is obese, falls asleep easily during the day and has an elevated level of blood carbon dioxide.
plasmapheresis - technique of separating out certain proteins from the plasma. Plasmapheresis is used to treat Guillain Barré syndrome and other illnesses.
pneumonia - infection of the lung tissues; can arise from many different types of micro-organisms, e.g. bacteria and viruses.
PO_2 - Partial pressure of oxygen (O_2) in the blood. Any value for PO_2 above 60 is usually considered a safe level; lower than 60 indicates hypoxemia and potential danger for the patient.
pressors - intravenous drugs used to support or raise a low blood pressure. One commonly used pressor is dopamine.
psychosis - severe mental disturbance characterized by personality disintegration or some loss of contact with reality; schizophrenia is one form of psychosis.
pulmonary - referring to the lungs.
pulmonary angiogram - dye is injected into the main pulmonary through a catheter, to outline the arteries within the lungs; this test is occasionally used to help diagnose pulmonary emboli.
pulmonary embolism - see emboli

respiration - general term for the process of bringing in oxygen from the atmosphere to the blood and excreting carbon dioxide from the blood to the atmosphere. Respiration is made possible by the process of breathing.
respirator - see ventilator.
respiratory failure - condition where the lungs have failed in their primary function of bringing adequate oxygen to the blood and of excreting carbon dioxide; in respiratory failure the level of blood oxygen is either reduced or the level of carbon dioxide is increased, or both.

scan - a general term for a variety of tests that "scan" or survey a part of the body, usually done in the radiology department. A lung scan is done to look for pulmonary emboli.
sepsis - infection involving the blood stream.
subcutaneous - under the skin; a subcutaneous injection is one given just under the skin, into the subcutaneous tissue.
surgical intensive care unit - area of hospital for patients who need intensive care after an operation (e.g., after heart surgery), or patients who have suffered major trauma (e.g., gunshot wound). (Compare with MICU, coronary care unit.)

tachycardia - fast heart rate, usually over 100 beats per minute.
TB - see tuberculosis.
teratogenic - able to cause birth defects.
trachea - medical name for `windpipe,' the airway that connects the back of the throat to the lungs. The trachea is the largest airway and divides into two bronchi.
tracheostomy - a surgical procedure that places a hole in the trachea, through which is inserted a short (usually plastic) tube. Tracheostomy is almost always done on patients who need long term artificial ventilation.
tuberculosis - disease caused by a bacteria called *mycobacteria tuberculosis*; abbreviated TB. TB usually involves the lungs but may also appear in any part of the body.

vasoconstrictor - any substance that constricts or narrows blood vessels.
vein - blood vessel that carries venous blood from the tissues back to the heart; venous blood is low in oxygen. See arterial.
venous - pertaining to the veins, e.g. venous blood.
ventilation - a general term for the physiologic process of delivering fresh air to the lungs for gas exchange. The term is sometimes used interchangeably with respiration.
ventilator - a machine capable of taking over a patient's breathing, also called a respirator. See artificial ventilation.

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