Disturbances of Bone Growth in a Child Who Survived Septic Shock

Louise S. Acheson, MD
Cleveland, Ohio

DR. LOUISE S. ACHESON *(Assistant Professor of Family Medicine):* Today's Family Practice Grand Rounds presents a case of a child with presumed Hemophilus influenzae sepsis at age 8 weeks, septic shock, and purpura fulminans, who at age 21 months presented with multiple skeletal deformities due to avascular necrosis of epiphyses, diagnosed as a late complication of septic shock. Her parents had obtained follow-up only sporadically for the child's problems and remained uncertain whether to comply now with the pediatric orthopedist's recommendations. In a meeting with me, their family physician, it became evident that the parents' religious beliefs, as well as their negative perception of events during the child's first hospitalization, played a large part in their decisions. Only by understanding these factors and the parents' explanations of the child's illness could the family physician mediate between this family and the medical care system to achieve continued care for the child.

THE FAMILY

The family consists of the parents, who have lived together for nine years and have been married for five, and their four children. They live in a crowded, four-room house, one of only a half-dozen houses on a highway bordering a large industrial complex. The father, G., aged 35 years, was born and raised in rural Tennessee. He recently quit his job as a shipfitter because of his religious beliefs. His wife, L., a homemaker aged 31 years, was born and raised in northern California, where her family still resides. Neither L. nor G. retains close ties with the families of origin. Their older daughter, A., aged 8 years, was born at home in California with a physician in attendance. Their son, D., aged 7 years, had an uncomplicated birth at home without any medical attendant. After the birth, L. was hospitalized with endometritis. Their daughter, J., aged 2 years 8 months, is the focus of this presentation. Another son, S., aged 1 year, was born at home with a family physician in attendance. Both parents, often accompanied by their small children, spend many hours a week visiting houses as Jehovah's Witnesses. For this presentation they consented to an interview, videotaped in their home, with the entire family present.

CASE PRESENTATION

I first met this family in June 1982, when L. was pregnant with her third child. L. was transferring care from a midwife to a physician, but still wanted to give birth outside the hospital. The pregnancy was uncomplicated. Labor began at 3:30 the morning before her due date. She came to the birth center three hours later. Her two children, husband, and a neighbor stayed with her throughout labor and the birth. At 7:01 AM, the membranes ruptured spontaneously, and the baby's heart rate dropped from 140 to 60 beats per minute. Two minutes later, an 8 lb 1 oz girl was born. Initially, her heart rate was less than 100 beats per minute and she did not breathe spontaneously. She received one positive pressure breath with bag and mask, and rapidly resumed normal heart rate, tone, and cry. The Apgar score was 5 at one minute and 10 at five minutes. J.'s physical examination was entirely normal, and her blood glucose level was greater than 40 mg/dL. The family went home together two hours after the birth. Mother and baby did well.

Later, I asked J.'s parents to describe their reasons for choosing an out-of-hospital birth:

L.: I have never had one in the hospital and I never want to . . . !

DR. ACHESON: Are there particular reasons you have in mind?

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From the Department of Family Medicine, Case Western Reserve University, Cleveland, Ohio. Requests for reprints should be addressed to Dr. Louise S. Acheson, Department of Family Medicine, University Hospitals Health Center, 2078 Abington Road, Cleveland, OH 44106.

L.: All I have to go on is what I've heard of other people having to go through: the nurses and the regulations. . . . It's not, 'What do you want to do?' It's, 'You're going to do this.'

DR. ACHESON: So a big thing for you is to have control over what you do or don't do.

G.: I don't feel that hospitals are healthy environments. They are full of sick people. A baby's got no business there!

DR. ACHESON: That brief problem that we had with J. didn't make you think differently about it for your next birth?

L.: No. I figure it's a once-in-a-lifetime situation.

DR. ACHESON: At the 10-day and 7-week checkups, everything was normal. Breastfed, J. was at the 25th percentile for height, 50th percentile for head circumference, and 75th percentile for weight. One week later, both parents had "colds." Eight-week-old J. awoke at 3:00 AM, vomited, and didn't nurse well. At 10:00 AM she seemed lethargic, moaned, would nurse for only one minute, and had had four watery stools. She had a fever to 38.5 °C. At 2:00 PM, I examined her in my office. She had a rectal temperature of 39.6 °C; her pulse was strong at 164 beats per minute. She was pale and listless, though she did nurse well after a dose of acetaminophen. She had a few pink papules on her chest and diaper area, but no petechiae or purpura. The rest of the examination was unrevealing. After speaking with a local pediatrician, I sent J. with her parents to the children's hospital, 15 miles away, for septic workup and admission.

By the time she arrived there and was seen, J. had vomited two times and had petechiae and diffuse purpuric lesions all over her body. Her temperature was 37.4 °C, respiratory rate 44, pulse 160 beats per minute but thready, her cry weak, and her blood pressure 48/0 mmHg. She was treated immediately with intravenous ampicillin, chloramphenicol, normal saline, and dexamethasone, bringing her blood pressure up to 100/0 mmHg. Her white blood cell count was $3.4 \times 10^3/\mu L$, with 11 percent polymorphonuclear cells and 18 percent band cells; her hematocrit was 28 percent. The cerebrospinal fluid contained 24 red cells and 1,500 white cells per milliliter, including 1,100 polymorphonuclear leukocytes; cerebrospinal fluid glucose concentration was 47 mg/dL, and protein 166 mg/dL. There was no growth of bacteria from blood or cerebrospinal fluid obtained after one dose of antibiotics. Three urine specimens were positive for Hemophilus influenzae antigens by counterimmunoelectrophoresis. A chest x-ray examination showed patchy streaks from the left hilum to the left lower lobe, consistent with either atelectasis or pneumonia.

Despite large volumes of fluid and dopamine, she continued in shock, with oliguria and fulminating purpura. The physicians had explained to J.'s parents that their baby needed albumin to treat shock, but they refused, for religious reasons, to consent. The physicians obtained a court order to administer albumin. Later, she required treatment for congestive heart failure with digitalis and diuretics.

DR. ACHESON: In the videotaped interview, I asked L. and G. to describe their experience at the hospital.

DR. ACHESON: At 8 weeks old, J. came in with a fever and got rushed to the hospital. Why don't you tell me in your own words what it was like for you when she went there?

L.: The worst part developed on the way there—she started getting petechiae. I didn't understand it.

DR. ACHESON: Did you see them and start getting really worried?

L.: Yes.

G.: I'd never seen petechiae before, and it was obviously something out of the ordinary and serious.

DR. ACHESON: What did they look like?

L.: Spots. They were just like purple spots. Her face, her arms, her body . . .

G.: Her entire flesh became patterned with it, just in the space of a few minutes.

DR. ACHESON: Was she still responding to you?

L.: Yes, she was responding, but she was very . . .

G.: Listless. Obviously didn't feel good.

L.: They took her right in as soon as they saw her. Of course, they knew we were coming. They had me take her in and lay her on the table in the emergency room and immediately rushed me out. I wasn't able to be with her, and that upset me very much. They didn't want me to know what was going on, so that I could be a witness to what was happening. They didn't want me there—thought I would be in the way.

DR. ACHESON: It doesn't sound as though there was much discussion about that.

G.: No. There was none.

DR. ACHESON: So, you and L. were there waiting.

L.: We waited in the waiting room, and periodically the doctor would come in and talk to us.

G.: They worked on her for awhile; then they came out with a consent form. It was then, you know, when we balked at the consent on the blood, that the excitement started. They'd already gone ahead and started giving her albumin before they discovered our religious preference.

DR. ACHESON: So, you're saying that they asked for your consent after they had already given albumin.

L.: Yes, after. Everybody was working on her at once. The first thing they did was to begin antibiotics.

G.: . . . which invalidated all of the tests that they took afterward, so they never really found out what was wrong with her.

DR. ACHESON: J. remained on ampicillin and chloramphenicol for two weeks. Her marrow was suppressed. Her hematocrit was 23 percent at the time of discharge and 35 percent one month later. J.'s purpuric skin lesions necrosed but did not require skin grafts. They were treated with silver sulfadiazine (Silvadene). She has deep scars with atrophy of underlying tissue scattered over her body, particularly on the flexor surfaces of her legs and arms. She underwent home physical therapy for about two months to prevent contractures.

Going back over that time, I tried to elicit what supports the G.'s had had. They stated that their main

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source of support was their involvement with Jehovah's Witnesses. As L. said, "It's a way of life, not just a religion! Every experience is a test to determine if you're true and faithful."

DR. ACHESON: Do Witnesses meet together in an organized way?

G.: Yes. We have five meetings a week. In addition to these meetings, we also go out on field service. That's the one that we're famous, or infamous, for where we knock on people's doors and wake them up on Saturday morning, or anytime. We do that every day of the week.

DR. ACHESON: How big is the group that meets together?

G.: There are 63 families in our congregation, which is the most intimate group. Even for people whom we've never met, however, if they are Jehovah's Witnesses, our homes and resources are completely at the disposal of our brothers. It's literally a closer-than-blood relationship.

DR. ACHESON: Can you recall any specific things that happened when your baby was in the hospital that were supportive and that helped you get through it?

G.: Well, the couple that was studying with us was there at the hospital with us for . . . how many days?

L.: Close to three days. There were many others, but this couple was with us all the time, so that they would be there to help if there was anything they could do.

G.: They did all my talking for me at the hearing. I was in no state to talk. I wasn't baptized at the time, I'd been studying for years, and I was barely able to restrain myself from violence. I told them I'd better not talk to these people or I'd hit somebody.

Table 1 is a time line of the G. family's medical care over the next 2½ years. As you can see, follow-up was sparse. I can best describe L.'s affect as bland. She would not discuss with me her feelings about the hospital stay. She continued to deny anything different about J. She used the Health Department, where J. was seen by a nurse, for routine care after one posthospital visit with me. She postponed follow-up at the Developmental Disabilities and Physical Therapy Clinics, but didn't say why.

When J. was one year old, in a very thorough examination at the Developmental Disabilities Clinic, no abnormality of her bones or joints was found. Her tooth enamel was defective, however, and this was attributed to the septic shock in the past. She was microcephalic, but appeared to be developing normally. Her mother became pregnant and received prenatal care in our practice, but J. was not seen. At 14 months, J. began to walk. Gradually, she developed a deformity of her left leg and shoulder. At 21 months, I saw her during her brother's visit for otitis media, and urgently referred her to a pediatric orthopedist because of a marked varus deformity of her left leg.

Physical examination revealed profound destructive changes involving her left shoulder, hips, knees, and ankles. There was limited range of motion, without pain, in the left shoulder, and a bony prominence attached anteriorly to the proximal humerus. There was

TABLE 1. TIME LINE OF MEDICAL CARE FOR THE INDEX PATIENT, J.

Date	Event
7/16/82	J. born
7/26/82	10-day checkup
9/7/82	Well-child checkup—7 weeks
9/13/82	Sepsis, shock, disseminated intravascular coagulation; court custody; transfusion; hospitalization
9/28/82	Home. Physical therapy
11/9/82	Follow-up. Growth delay, decreased head growth; immunized at health department
2/23/83	Seen by family physician (brother's visit)
	Physical therapy and developmental evaluation
7/25/83	14 mo. Seen in Developmental Disabilities Clinic: normal mentally, dental problems; failed dental clinic follow-up. Immunized at health department
	L. pregnant
1/12/84	S. born at home
4/9/84	21 mo. Family physician noted leg deformity when J. accompanied her brother to an office visit
5/16/84	Orthopedic clinic evaluation and x-ray examination
7/26/84	Family meeting with family physician
10/84	L. and G. baptized

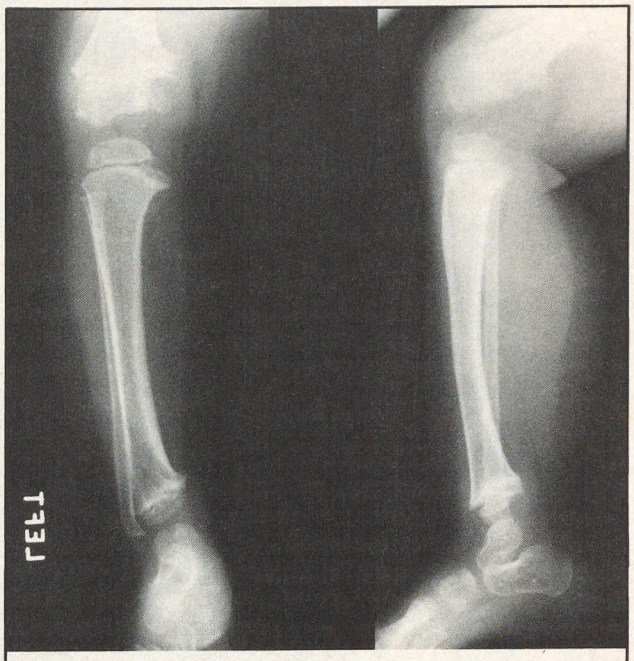

Figure 1. Anteroposterior and lateral x-ray films of the patient's left leg at 22 months

crepitus in both hips, and limitation of external rotation to 70 degrees. There was a 35-degree varus deformity with significant laxity of the left knee. The left ankle assumed a varus position at rest. X-ray films showed marked destruction of the left humeral head, which had fallen into a marked varus position. Destructive changes were seen in both femoral heads, the left knee, and the ankle (Figure 1). Destruction of the left medial femoral condyle appeared to be the cause of the varus deformity at the knee. In the ankle, the epiphysis of the distal medial tibia had closed prematurely, resulting in a varus deformity. The left femur was 0.4-cm shorter than the right, and the left tibia and fibula 1.0 cm shorter than the right (Figure 2).

The destructive changes seen in J.'s bones are consistent with avascular necrosis of multiple epiphyses. Similar disturbances of bone growth after sepsis with disseminated intravascular coagulation in infancy were first reported in 1981.[1,2] Previously, few children survived such an illness. Two types of lesion characterize this disorder[3]: (1) asymmetrical destruction of major epiphyses with disturbed growth adjacent, and (2) multiple, rather symmetrical lesions of growth plates in the lower extremities, particularly affecting the central part of the epiphyseal cartilage, leading to "cupped" epiphyses in the following manner. Epiphyseal plates are supplied by end-arteries. With decreased perfusion the central growth plate may suffer ischemic damage, while the margins, receiving collateral circulation from the periosteum, survive. Subsequently, the edges of the epiphysis may grow, while the center does not; eventually, a central bony bridge may form, precluding further growth. The results of such lesions are bowing and asymmetric growth of extremities, shortened bones and resulting dwarfism, and deterioration of weight-bearing joints. All reported children with such profound and widespread damage were less than 1 year of age at the time of sepsis. All suffered disseminated intravascular coagulation and septic shock with meningococcemia. This patient's illness was similar to those previously reported, but antibody studies suggest that she had Hemophilus influenzae sepsis.

Treatment of such a recently described entity is fraught with uncertainty because the growth potential of epiphyses damaged in infancy by ischemia is unknown. In the hope of maintaining growth, surgeons may excise the bony bridge, interposing fat or silastic. Wedge osteotomies may be needed to correct angular deformities. Leg length discrepancies may be treated with a heel lift up to 6 cm in thickness. Surgery is best done after 10 years of age but while the patient is still growing. A combination of techniques may be used to limit the growth of the longer leg (epiphysiodesis) and to lengthen the shorter one (eg, the Wagner procedure for osteotomy and distraction with bone grafting).[4] Joint replacement may be needed if weight-bearing joints are destroyed. The first step in management for J. will be careful measurements of bone growth by serial x-ray studies. Excision of a bony bridge in the left knee was recently recommended.

The parents are now trying to decide on treatment for their daughter. L. remarked that it would be easier if J. could decide for herself. At a family meeting more

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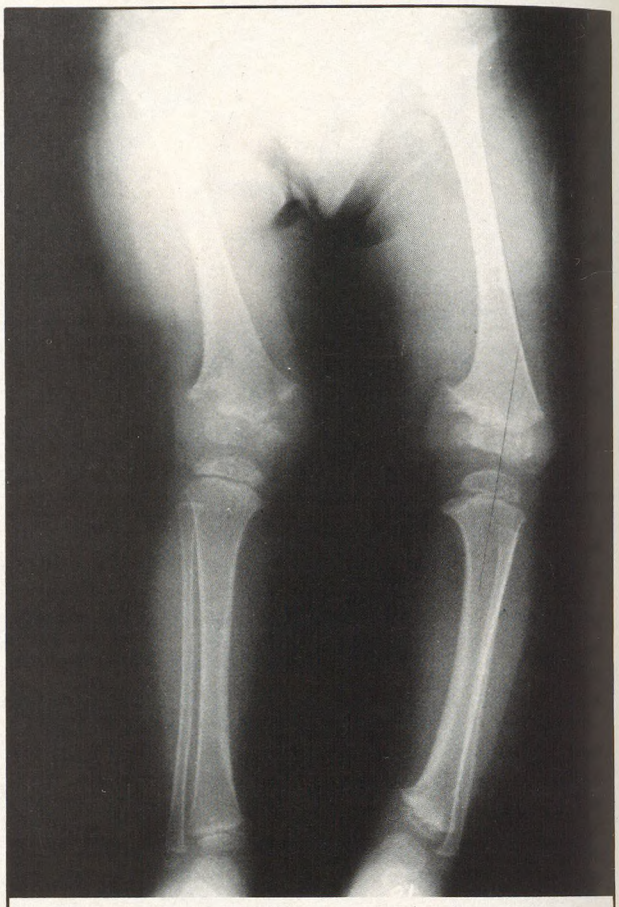

Figure 2. Anteroposterior x-ray film of both lower extremities at 22 months

than two years after J.'s original illness, they revealed to me their feelings about events during her hospitalization, and the extent to which this has influenced their use of medical care.

DR. ACHESON: Did your experience with the hospital affect the kind of medical care you got for her after that?

G.: Yes, definitely. We are frightened. I have been impressed at how much more credence the legal system gives to a doctor's word than they do to anything else. We've been convinced! They showed us the power! They took custody away from us: 'Not your baby anymore. We'll do what we want with it!' I'm very leery of even going around a hospital with any of my babies. I try to remain rational enough to obtain what I personally deem to be necessary medical attention for my children, but that feeling is there, nonetheless.

L.: Our child could get taken away from us if we don't go according to what they want. If we would have gone strongly against them any further, they could have just put her in another home.

DR. ACHESON: How about more subtle ways? Has it affected the kind of medical care you get for her, other than in hospitals, for example? Are you more cautious about what you would do for her if she gets sick?

L.: Well, so far, she's just been normal. I've been taking her for x-ray examinations, and that in itself is scary to me

because of what they want to do.

G.: We're afraid to talk. We never know whether something we say is going to strike someone wrong, and they'll decide it's in their best interest to take matters into their own hands.

DR. ACHESON: Are you afraid to ask questions about what's recommended or about what could happen to her?

L.: We ask them questions, and they tell us what they think. They beat around the bush a lot. It's as if they see, instead of a general, overall [plan], whatever is the worst at the time.

G.: Whatever their field of specialty is.

L.: First, I took her to a foot doctor.

G.: He wants to work on her ankle, and isn't concerned about anything else.

L.: And then we took her to the leg doctor, and he wants to work on her knee.

G.: It's too fragmented. We can't really discuss her as a whole human being.

DR. ACHESON: If you were discussing her as a whole human being, what kinds of things would be considerations in your mind?

L.: Let time tell. Personally, I would just as soon she not have to go through pain. If she's going to have to be in pain from the way she is, I would rather they try to do something to straighten her if they can, which is still an if, that is, what they can do.

G.: The prognosis is not good right now. The tomograms reveal some bridging on the growth plates. However, the doctors don't really think there's much chance of improvement if they go in there and burr them out. If the bridging had been due to some trauma, rather than an infection, there'd be a greater chance of therapy being effective and her bones correcting through growth.

DR. ACHESON: What do you understand what she'd be like as she grows up?

G.: When she grows up, my prognosis is that she'll be healthy. According to our religious beliefs, the interim is the only thing that we're concerned about (because within the next few years the millenium will come, at which time everyone's body will become healthy and whole). This is where we differ with the doctors. They feel that what they have to offer is all that can be done for her for the rest of her life, and we don't see things that way.

DR. ACHESON: Even the way you see it, you may be dealing with a period of 10 or 15 years.

G.: My feelings on it are that, since they don't forsee being able to do much, even with all these repeated surgical procedures, I'd just as soon do without them, unless it comes to a point where she is no longer ambulatory. If she's going to be in a wheelchair, then we should attempt to correct it surgically.

L.: My understanding of bone surgery is that it's very painful and hurts for a long, long time. Bone surgery isn't like any operation that would be over with in a short time. I'd be weighing it, balancing it out, trying to determine whether the pain she's got to go through is worth what she's going to get out of it. And you can't get doctors to help you with that!

G.: We really can't tell at this point. She's not handicapped at this point; it doesn't stop her from doing anything at all. We don't know what handicapping will occur as she grows. We're just playing it by ear, which is actually the same thing that the doctors want to do.

DR. ACHESON: The other thing I'm interested in is the effect that this chronic problem has on J. as a person. Does she already know? Does she realize that her body is different from other kids?

L.: She's just starting to. I've seen her looking at her leg . . . feeling her bones and whatnot. I think she's beginning to realize there's something wrong. She's had it as long as she knows.

DR. ACHESON: This family displayed suspicion and fear of the medical care system. The couple was united in using denial and projection as defenses to cope with their child's serious illness. Their beliefs and participation in a religious group provided strong reinforcement for their attitude toward the medical system, their explanation of the child's illness, and their view of the prognosis. Particularly striking was their conviction that the millenium would come during J.'s childhood, and then her body would be healed.

The family's behavior raised many concerns for me as their physician. As many physicians must, I had guilt feelings about inadequate follow-up of a "noncompliant" family. To this was added the parents' legitimate fear that, should they refuse treatment for their daughter, they might lose custody of her, and their desire to avoid such a circumstance. It is difficult for me to determine to what extent I should present myself as separate from the medical care system at which these parents were angry, or to what extent I should defend or explain the system to them. The family's beliefs tended to polarize the issues; my task was to find a common ground. Together, we had to tolerate uncertainty about the benefits of treatment and long-term prognosis, and yet make a plan. With knowledge of the family's beliefs and coping style, a family physician may be in a special position to mediate between the family and the medical care system for a better outcome. In this case, the outcome is still an open question.

This child's bone-growth abnormalities exemplify a newly reported disorder, a late sequela of septic shock with disseminated intravascular coagulation in infancy. This case is the first reported of widespread epiphyseal damage associated with Hemophilus influenzae sepsis; previously reported cases were associated with meningococcemia. Long-term follow-up may provide needed information about the growth potential of bones thus damaged.

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