shocks. Precordial compression is held in abeyance during this interval. Yet experimental evidence in our laboratory indicate that if precordial compression is withheld for an interval of as little as 30 s, without continuing precordial compression, the likelihood of successful defibrillation and survival is remote. Accordingly, there is a great need to reinvestigate this problem.

The resolution of this and so many other issues that pertain to CPR is likely to be achieved by ensuring careful and objective scientific research rather than consensus in the absence of secure data. Hopefully, these experiences will therefore prompt a reawakening of the need for such research and its funding. After all, it relates to the survival of no fewer than 400,000 victims of out-of-hospital cardiac arrest in the United States alone in each year.

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Death by Syrinx

Worse Than Ondine’s Curse?

Sleep disordered breathing is relatively common in the population, with sleep apnea syndrome (SAS) representing the most serious manifestation. The natural history of SAS is largely unknown, and although degrees of pathophysiology exist (severe, moderate, mild), there is little evidence showing that progression from health to severe disease occurs according to these categories. Few would argue, however, that diseases linked to Ondine’s curse represent singular disorders associated with predictable fatal central apnea if not identified. After our patient died of syringomyelia (SM)/syringobulbia (SB), we reviewed the literature, and we acknowledge that this rare disease is equally capable of causing premature respiratory death. This uncommon lesion of the central nervous system is unfamiliar to most pulmonologists. Indeed, a recent comprehensive review on SAS fails to include SM/SB, and a chapter in a neurology textbook also does not make the association of SM/SB with symptomatic airway patency problems. Accordingly, in view of its lethality, it should receive the recognition, and respect, equally afforded to Ondine’s curse.

Although Estienne was the first to describe cavitarias in the spinal cord, and Ollivier D’Angers coined the term “syringomyelia”, the brave Roman Mucius Scaevola was probably the first recorded person suffering from the malady when demonstrating his fearlessness to the king of the Etruscans, Porsenna, he put his hand into a blaze (Mueller quoted by Bodechtel and Schrader10). The etiology and pathogenesis of the disease is largely unknown, although syrinx cavities have now been described in up to 22% of posttraumatic injuries and have been associated with Arnold-Chiari malformation. The important pathologic change of syringomyelia is a gliosis that precedes cyst formation. The growing cavity occupies the central portion of the spinal cord, most frequently in the cervical region. It may extend laterally into the gray matter of the anterior and posterior horns of the spinal cord (syringomyelia) or cephalad into the lateral tegumentum of the medulla oblongata (syringobulbia).

The usual symptoms of an invading syrinx are the loss of pain and temperature sensation, with preservation of the sense of touch, position, and vibration. The intimate anatomic location of a syrinx to the central nuclei of respiration and the cranial nerves presents an ominous threat to normal breathing. Respiratory abnormalities are serious and include hoarseness, aspiration, dysphonia, and stridor. Diaphragmatic weakness and unilateral and bilateral vocal cord paralysis have been described also. Furthermore, hyperventilation syndrome, apnea, and sudden death are additional respiratory hazards of this disorder. Of note is the fact that of the 12 patients described in the literature with SM/SB, 5 died suddenly.

In view of the number of respiratory disorders that SM/SB are associated with, and the myriad of more common entities equally capable of causing sleep apnea syndrome, SM/SB can be easily overlooked.
Accordingly, it seems prudent that a very careful history be obtained (especially for spinal cord trauma) and a thorough neurologic examination be performed in all patients with airway patency problems of obscure etiology. The clinical neurologic picture is so characteristic that diagnosis thereafter is seldom in doubt.6 Contrast myelography and delayed CT scanning performed with the patient in the supine, head-down position, will usually expose the syrinx. Magnetic resonance imaging can now demonstrate spectacular views of a syrinx in the sagittal plane of the brain and spinal cord.5,7

Management of SAS secondary to SM or SB rests on the nature of the airway patency problem. These techniques include CPAP masks, negative pressure ventilation, positive pressure pneumobelt, tracheostomy, and continuous nocturnal mechanical ventilation.4,15,18 The only specific therapy for SM/SB is surgical decompression of the foramen magnum and upper cervical canal.6 These procedures, however, have varying results and are largely unpredictable.

Unlike acquired Ondine’s curse, which may have permanent, but stable disease,19 SM/SB tends to be a progressive disorder. Although SAS symptoms may be managed with conventional techniques, effective intervention to limit the growth of the syrinx is not presently available. Accordingly, SM/SB may represent a universally fatal respiratory disease once airway symptoms are noticed.

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Tough Love and the Pulmonologist

Marvin Olasky’s book, The Tragedy of American Compassion, has created a great stir in Congress. Evidently, the book has had a great influence on Newt Gingrich and other conservative congressmen. It has fueled the welfare debate by questioning the nature of true compassion. This historic treatise on the care of the poor reviews the position of government from the Puritan period to the present. It reminds us that before the late 1800s, true compassion was thought to be what is called today “tough love.” Laws were passed that forbade government and even private citizens from offering charitable assistance to drunks and others who were characterized by reprobate behavior. A willingness to change behavior was the requirement for charity from either private or public institutions. Currently, we provide Social Security disability benefits to individuals who are confirmed alcoholics and drug abusers. Our forefathers would say that this reinforces self-destructive behavior.

What about the pulmonologist who treats the patient with cigarette-related disease such as resectable lung cancer or COPD? How often are we asked so subtly to “fix my problem so I can continue to enjoy my smoking habit?” Refusing to play this game is met with cries and accusations that we are uncaring and uncompromising physicians. However, now the shoe is changing to the other foot, as medicine plunges headfirst into managed care. Congress and health-care economists