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Medicine & Health Rhode Island

VOLUME 84, NO. 12 DECEMBER, 2001

PUBLICATION OF THE RHODE ISLAND MEDICAL SOCIETY

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Medicine and Health\Rhode Island *(USPS 464-820), a monthly publication, is owned and published by the Rhode Island Medical Society, 106 Francis Street, Providence, RI 02903, Phone: 401-331-3207. Single copies $5.00, individual subscriptions $50.00 per year, and $100 per year for institutional subscriptions. Published articles represent opinions of the authors and do not necessarily reflect the official policy of the Rhode Island Medical Society, unless clearly specified. Advertisements do not imply sponsorship or endorsement by the Rhode Island Medical Society. Periodicals postage paid at Providence, Rhode Island. ISSN 1086-5462. POSTMASTER: Send address changes to Medicine and Health\Rhode Island, 106 Francis Street, Providence, RI 02903. Classified Information: RI Medical Journal Marketing Department, 175 Mathewson Street, Providence, RI 02903, phone: (401) 331-4637, fax: (401) 331-3594*

COMMENTARIES

Move Over Lance Armstrong

I am often struck by how well many of my patients deal with their **Parkinson's disease (PD)**. I am sure they do better than I would in their shoes. The American Parkinson Disease association has the motto, "Accommodation Without Surrender," which is as good a motto as I've come across. Certainly PD poses limitations but it's not a life-ending diagnosis. I may be wrong but I've developed the belief that PD patients who are intermittently disabled by severe clinical fluctuations, but able to walk independently at some time while completely immobile at others, are much harder on themselves, less accepting of their situation than patients with fixed deficits, such as post - traumatic paraplegics who have no realistic hopes of walking again. It seems to be harder to accommodate when you don't always have to and most PD patients fluctuate to some degree, with a wider amplitude than those with enduring, stable neurologic impairments.

Lance Armstrong is, of course, the super-talented bicycle racer who has won the Tour de France three times, most recently after completing chemotherapy for metastatic testicular cancer. His success after chemo has made him a super hero, a guy with "heart," who doesn't give up. I am less impressed. Lance Armstrong is one of the great athletes of all time and, presumably, likes doing what he does and likes being the best in the world. Why would he not return to competition after chemotherapy? Certainly he'd be weaker from the inactivity and the medication effects, but why would he not return to doing what he does so well, especially when it became clear how well be could do it after his treatment.

In contrast, look at Don Brennan.* He is a 50-year-old man who has had PD for four years. He competed in triathlons before his PD emerged and continues to participate despite his PD. He completed the Iron Man Triathlon in Hawaii last year (2 miles of swimming in the ocean followed by a 110 mile bicycle race around the island, followed by a full marathon road race), which he finished in less than the 17 hour official cut-off time. Aside from slowing down, his other accommodation to PD was his inability to pick up water cups while riding his bicycle. He had to stop his bicycle but when he did this, his tremors worsened and the race monitors tried to pry him from his bicycle and get the medics to care for him. "Leave me alone. I have Parkinson's disease. I'm feeling great." That was last year. Recently he completed a triathlon at Lake Placid and he's entering another in British Columbia.

I exercise daily but I have never entered a triathlon. I certainly could never have completed a "real" triathlon, maybe one of the mini-triathlons with markedly reduced distances. When I was younger I aimed to go faster and farther, albeit never "fast." I then reached a point at which the goal became maintenance rather than improvement. And now I hope to reduce the accelerating decline I'm currently experiencing. I am slower. This is not a figment of my imagination. I take longer to run a fixed distance. I swim fewer laps in a fixed interval. On the one hand this is frustrating but I tell myself I'm just trying to be healthy. If I didn't exercise I'd be slower, heavier, with higher cholesterol, higher blood pressure and a faster heartbeat. I tell myself that if I can preach to my patients the mantra of 30 minutes exercise per day, then I should follow my own advice. Why should anyone follow me if I don't lead? Then I think about Don Brennan. How does he spend 3-4 hours per day, after a full workday, in training? Grueling training. Not only does the PD slow him down, but also it worsens the post-exercise fatigue and delays the recuperation. And then what are his goals? He's losing ground on his old compatriots. While they've all aged equally, the rest of them have normal dopamine levels. He can't be looking for any records. He's not interested in being the first Guinness Book of Records entrant with PD. He's doing this for himself, just like most people who climb mountains, run marathons or swim the Bay. But, unlike most of us, he is unfazed by his need to accommodate. "Oh yeah, I'm slower than I used to be. I have to stop my riding to pick up my water cup." It is almost as if he is saying, "I'll take your best shot. Nothing is going to get in my way."

Don leaves me in the dust. I often wonder how well I'd stand up to PD. Would I be a portrait in courage like Don, like the medical school dean who announces at the onset of each new class that be has PD, not to pay attention to his movements, or the corporate vice president whose job it is to tell his sales personnel to smile more, explaining, of course, that his own facial expression is masked by PD; or the 38 year old woman who had her fourth child while suffering from PD and now works half time with two small kids. Hardly your "ordinary heroes."

Lance Armstrong is very impressive. He didn't give up. He overcame adversity. He, however, was cured. While he lives with the sword of Damocles over his head, that he may suffer a relapse, which he can do nothing to forestall, he is normal now. He had to choose between forging a new path and overcoming a hurdle to resume his former life. He put himself on the line, going all out to surmount an obstacle of unknown height. How different this is than living with a process that slowly but relentlessly lowers your skills, by aging your motor system before its time.

If medals for courage are to be given I'd give them to my patients. Their resolution is a constant source of inspiration. They make me work harder at work and help me get through my minuscule 30-minute workout each day. My muscles ache. I don't get enough sleep. I'm worse each week. Complaints? I don't have any. My admiration for my courageous patients increases my dedication. Lance Armstrong is quite impressive but our crusaders who fight their battles every day could teach him a thing or two.

– Joseph H. Friedman, MD

**Mr. Brennan gave permission for use of his name*

A Doctor's Rescue and a Lawyer's Poem

To declare that a practicing physician had been responsible for our national anthem sounds like an exaggeration if not a brazen presumption. Yet a sober review of the events preceding the composition of "The Star Spangled Banner" confirms that a Maryland physician named William Beanes did play an indirect but decisive role in its creation. History does not deny that if this physician had not been enjoying his dinner at home in Upper Marlboro, Maryland, on an early August evening of 1814, our national anthem would not have been authored by a Georgetown lawyer named Francis Scott Key.

The War of 1812 had been fought in a desultory manner until the British, incensed by the deliberate burning of the Canadian city of London Town [Toronto] by American forces, mounted a major invasion of the Chesapeake Bay region. Seaborne forces under the command of Vice Admiral Sir George Cockburn entered the Bay and proceeded up the Patuxent River. About 5,000 British and mercenary soldiers, under the immediate command of Major General Robert Ross, were then placed ashore at the tobacco port of Benedict; they proceeded north in the direction of Washington, paused at the town of Upper Marlboro, Maryland, and briefly occupied the home of the local physician, Dr. William Beanes.

The British expeditionary force continued its march toward the Capitol on the following morning, encountering only ineffective American resistance in the town of Bladensburg just south of Washington. Then, with little pause, they swept into Washington, burning the various government buildings, including the President's House [later to be called the White House] and the Capitol building. The British had neither the logistic support nor the hospital facilities for an extended occupation. Accordingly they retreated back to the west shore of Chesapeake Bay and boarded their ships in preparation for a major assault upon the city of Baltimore at the northern extremity of the Bay. The British troops wounded in the Bladensburg engagement were abandoned.

In the retreat from Washington, some straggling British soldiers, said to be drunk, lingered in Upper Marlboro for purposes of looting the local homes; a few invaded the property of Dr. Beanes, who was then at dinner. He promptly disarmed them; and, with the help of neighbors, had the British looters remitted to a jail in Queen Anne, a few miles north of Upper Marlboro.

Beanes, it should be emphasized, was a well-respected member of the eastern Maryland community. He was a third-generation American, a major land-holder and a trustee of St. John's College in Annapolis, Maryland. He had learned his medical skills through apprenticeship, serving with distinction as a surgeon in the army of George Washington during the battles of Germantown and Brandywine.

One of the British soldiers held in the Queen Anne jail managed to escape and return to the main body of British troops. His arrest, and the continued imprisonment of his fellow stragglers, incensed the British commander, who then sent a detachment of dragoons to take Dr. Beanes into custody. They arrived at the Beane mansion on the late night of August 29, awakened the doctor and took him prisoner with such speed, according to one story, that he failed to retrieve his eyeglasses. Beanes was then placed in solitary confinement aboard one of the British warships with the expectation that he would stand trial in Halifax.

Beanes' friends, alarmed by the turn of events, appealed to a Georgetown lawyer named Francis Scott Key, a graduate of St. John's College and a friend of Beanes.

Key then spoke with President Madison, temporarily quartered in northern Virginia. Madison, recognizing the violation of a signed agreement between Britain and the United States forbidding the imprisonment of civilians, authorized Key to visit the British commander, under a flag of truce, to arrange for the release of Dr. Beanes.

Accompanied by Colonel John Skinner, the American officer in charge of prisoner exchange, Key proceeded to Chesapeake Bay, boarded a small vessel and sailed to meet the Admiral's flagship to negotiate for Beanes' release. Admiral Cockburn initially refused; but when he learned that the British wounded back in Bladensburg were being adequately care for, he relented and authorized the release of the doctor. However, since his fleet was about to attack the city of Baltimore, he insisted that Key, Skinner and Beanes be temporarily confined to to one of the British ships until the naval engagement against Baltimore's fortifications had been completed.

Key, Skinner and Beanes witnessed the naval attack upon Fort McHenry, the principal guardian of Baltimore's shoreline. Since the defending Americans had intentionally sunk some vessels in the harbor, Cockburn's ships could not approach the Fort within the range of their guns; instead, they unleashed a late-night barrage of Congreve rockets which illuminated the skies over the fort and deafened the anxious observers on deck of the British sloop; but in truth did little damage to the Fort.

The sun rose on the morning of September 14, the dawn mists dissipated, and the fort had clearly withstood the intense British bombardment. Dr. Beanes, deprived of his spectacles, asked Key: "Oh, say, do you see whether the flag still waves over the Fort?" Key, in possession of a spy glass, saw the American flag [some 12 by 15 feet in size] still waving over the McHenry ramparts. Moved by this scene, and in response to the doctor's inquiry, Key - a lawyer by profession but poet by avocation - scribbled a four-stanza poem which he entitled, "The Defense of Fort McHenry."

The 65 year-old Beanes was eventually released to resume his Maryland practice of surgery. Key returned to the District of Columbia as its district attorney; and his hastily composed tribute to the defense of Baltimore was published within weeks, set to the melody of a British drinking song called, "Anaceron in Heaven." And General Ross? He was killed by a sharpshooter's bullet during the unsuccessful land attack on Baltimore. The British then retreated to their ships and the inconclusive war formally concluded with a treaty signed in Ghent, Belgium.

There are those, motivated more by musical standards than by patriotism, who would have wished that the gentle Dr. Beanes had not taken it upon himself to have British looters arrested, thus keeping Francis Scott Key from venturing forth to free him; and, by indirection, then allowing this nation to choose a more suitable national anthem.

– Stanley M. Aronson, MD, MPH

This issue of Medicine & Health/RI *is supported by unrestricted educational grants from Coordinated Health Partners/Blue Chip, Synthes Maxillofacial, and Leibinger Stryker Corporation*

At Face Value: Comprehensive Care for Problems Related to the Face and Calvarium

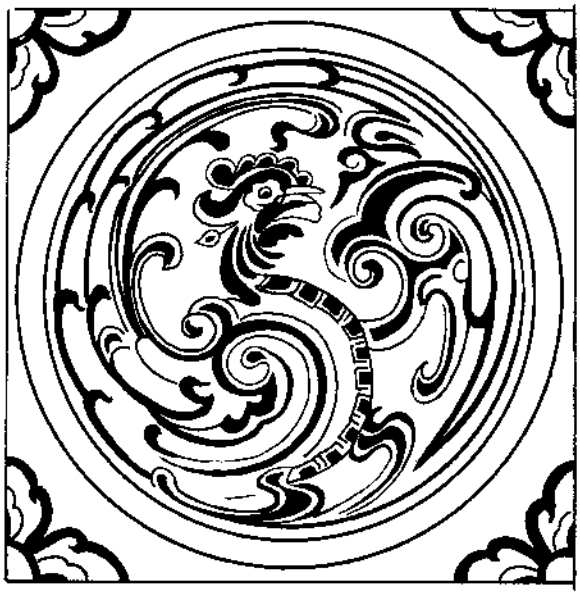
Patrick K. Sullivan, MD

Patients receiving plastic surgery care in Rhode Island benefit from a great deal of local expertise. A significant effort has been made to coordinate the comprehensive care given to patients with problems related to the face and calvarium. The primary care specialist is, of course, the cornerstone in the patient's care. Yet, due to the complexity of the problems, the treatment has become highly specialized and involves numerous other specialties and subspecialties. This is why the team approach has become so important.

It is not uncommon to have three different specialties involved in treatment planning and operating on the same child during one procedure at Hasbro Children's Hospital. A number of other team members who are not surgeons also are involved with the patient and are critically important in providing care. Early medical intervention, especially among children, has proven to facilitate excellent outcomes. And while we often focus on surgery, we find it can often be avoided if our smaller patients are referred at the appropriate time. This is especially true with a majority of head shape abnormalities.

This edition of *Medicine & Health/Rhode Island* features contributions made by closely associated specialties and subspecialties that focus on face and calvarial deformities. Our initiative is to provide readers of all specialties with deeper insight into other specialty issues and thereby improve overall patient care and the coordination of services. The objective of this collection of articles is to increase the sensitivity and awareness of primary-care specialists and other related specialties to the importance of optimal timing of treatment, because this is such a critical issue with growing and developing patients.

The complexity of treatment has given rise to the involvement of a number of specialists in the care of patients with problems related to the face and calvarium. And the overall current expertise in Rhode Island for treating complex facial and calvarial problems is impressive. A portion of this team effort is represented in this journal. Articles on head shape abnormalities and craniosynostosis are included from both a pediatric neurosurgical prospective and also a plastic surgical perspective. The approach to craniofacial pathology, from both pediatric otolaryngology and neurotology perspectives, also are included. Pediatric anesthesiologists discuss intra-operative and peri-operative special care, while hearing and speech issues are presented from a long-term perspective in Rhode Island.



Pediatricians in Southern New England practice their specialty at a very high level; some have had a particularly close relationship with our craniofacial team. Genetic counseling has proven very beneficial for our patients, especially now with our increased level of diagnostic sophistication. Experts in the area of molding helmets and conservative cranial vault shaping too have worked closely with us for years to provide wonderful results for selected children, thus elmininating the need for some surgeries.

Craniofacial and maxillofacial fellowship-trained surgeons frequently specialize in various aspects of treatment of complex craniomaxillofacial problems, as it is difficult for any one person to be expert in every aspect of treatment. We often find that maxillary and mandibular abnormalities are frequently part of craniomaxillofacial pathology, especially with syndromic patients. And in these cases, dental specialists prove particularly important team members as they assist children in correcting their teeth and occlusion.

Both clinical and laboratory research abound in Rhode Island as an effort is being made to advance the field. Additional updates are planned in *Medicine & Health/Rhode Island* to ensure a coordinated effort throughout the state and region in the care of growing, developing children and adolescents with abnormalities of the face and cranium.

Patrick K. Sullivan, MD, is Associate Professor, Plastic Surgery, Brown Medical School.

Correspondence:
Patrick K. Sullivan, MD
235 Plain St., Suite 502
Providence, RI 02905
phone: (401) 831-8300
fax: (401) 831-8304
e-mail: cosmetic@drsullivan.com

Artistic Shaping of Key Facial Features in Children and Adolescents

Patrick K. Sullivan, MD, and David P. Singer, MD

Shape and form often define art. Similarly, elements of facial and cranial shape are closely associated and bring form to the human face - our interface with the world. Key facial features are of primary importance in the development of the face, the person, and the personality. Structural elements such the nose, chin, and ears made up of bone and or cartilage are not only dominant features but also can be artistically shaped and sculpted to bring the face into a more normal, aesthetically pleasing balance.

Since the popularization of the term "plastique" via the *Handbuch der plastischen Chirurgie* in 1838, the discipline of plastic surgery continues to evolve.[1] The reconstructive aspects of the field in the pediatric population are widely recognized, particularly in facial dysmorphologies including the treatment of craniosynostosis. In contrast, the facial aesthetics of the pediatric patient with more subtle dysmorphology has often been less emphasized. The importance of facial aesthetics in the development of the personality, social skills, and self-esteem has been less estimated. Overall, conservative facial reshaping with a respect for the pediatric patient can have significant impact on children and adolescents.

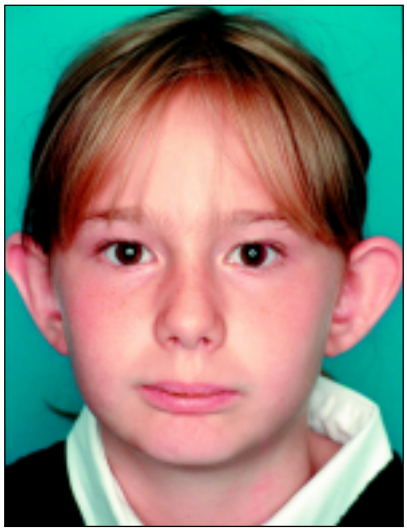


Figure 1a. The face pre-op.

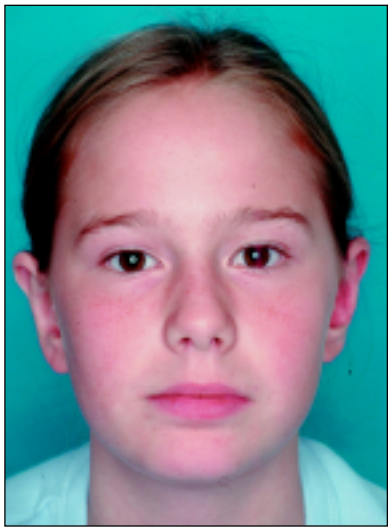


Figure 1b. The face post-op.

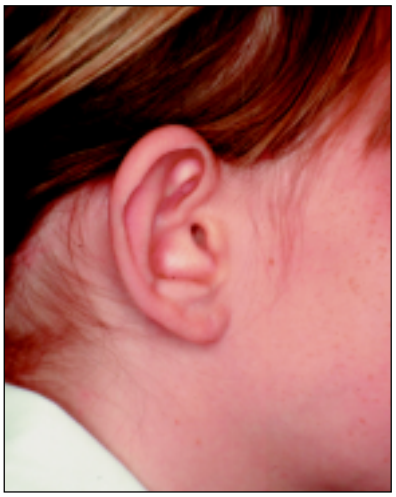


Figure 1c. The ear pre-op.

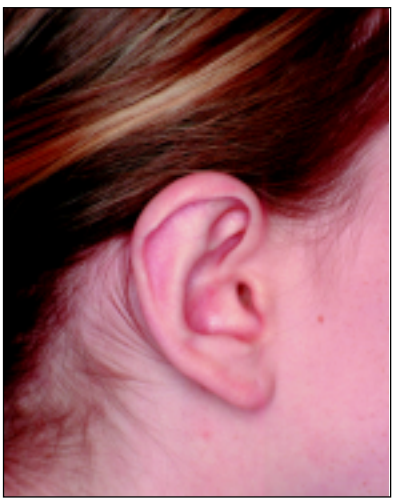


Figure 1d. The ear post-op, with the incision hidden behiond the ear.

Psychological influences define the perception of body image and underscore the way external appearance affects behavior. Plastic surgery to correct an unattractive feature can be enormously successful and remarkably free of conflict in this population. Children and adolescents undergo rapid reorganization of the self-image after plastic surgery with subsequent positive changes in behavior and interpersonal interactions.[2] Parents have reported to us dramatic and rapid improvements in the behavior of children who have undergone ear reshaping, for example. The ridicule of peers can be powerful. Adolescents with nasal deformities may curtail their socialization rather then deal with painful interactions and rejection. Patient selection and timing of surgery, however, are both important factors in achieving successful outcomes in these young patients. The core values to guide in patient selection for these procedures are physical development, age, psychological stability and maturity, and clear realistic expectations of both the patient and the family. Amongst the pediatric population, the areas of most common concern are the ear, nose, and chin.

Ear

The protruding ear, defined by prominence of the auricle beyond 1.8-2.0 centimeters from the helical rim to the scalp, can be a common finding.[3] The acceptable variability of ear prominence is dictated by individual preferences. The condition results from embryonic arrest during the final convolutions of the ear with failure of folding of the antihelix as well as overdevelopment of the conchal wall. [Figure 1] The deformity is usually found bilaterally.[4,6] Further, familial inheritance patterns have been observed, including an autosomal dominant expression pattern seen in the Caucasian population.

The anatomic manifestations and

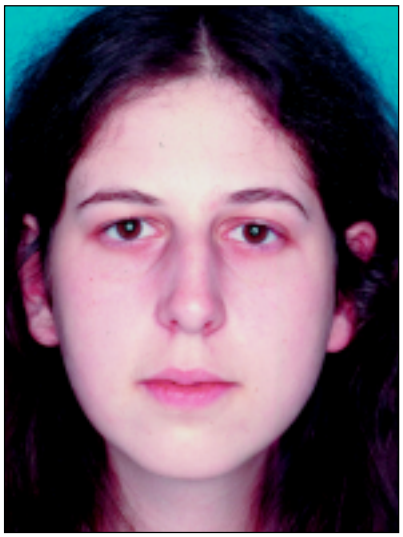


Figure 2a. This 17-year-old was concerned about the appearance of her nose. She did not like the nasal shape, especially the nasal hump.

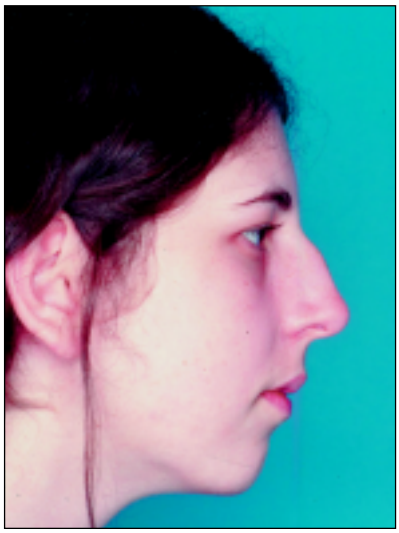


Figure 3a. Upon reviewing the photographs with the patient and her parents we also discussed the artistic facial balance and the chin position. We then went over treatment options to balance the face.

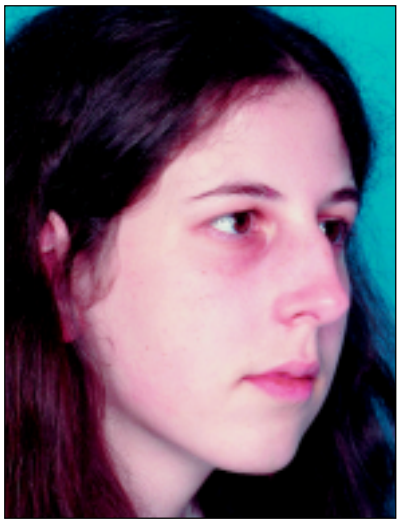


Figure 4A. The oblique view also demonstrates the retruded chin.

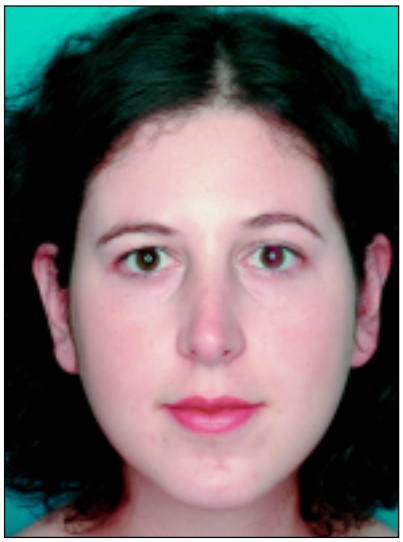


Figure 2B. The post-operative view five years after treatment.

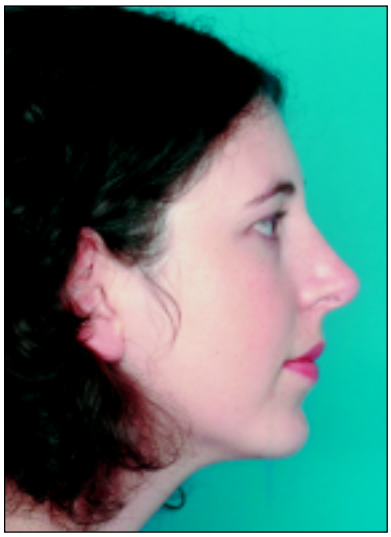


Figure 3B. The lateral view demonstrates how the dorsal nasal deformity has been removed and the nose has been reshaped all via internal nasal incisions. The chin advancement was done by an intraoral incision and an osteotomy, sliding the chin forward, where it was then fixed in a new position to give a better artistic balance to the face.

Figure 4B. The oblique view demonstrates the new facial balance.

distortions of prominent ears can be disturbing and distinguishing features for a child and his or her family. The appropriate time to address these concerns is governed by psychologic and physical development. The body image usually begins to form around the age of 4 to 5 years. At this age the child begins to be aware of being different and is motivated to conform. When this awareness begins, it will make him or her more cooperative with the surgery and the restrictions it entails.[4] Also, when the child starts school, s/he has increasing social interactions - again provoking self-awareness. On the other hand, a large percentage of the ear growth is not complete until approximately the age of 6. [5] The ear continues to enlarge mostly in its vertical dimension throughout life, predominantly due to elongation of the earlobe.[3] Hence, correction of the deformity via otoplasty is offered to patients at approximately age 6. Lastly, there is some credence to early tape repositioning of prominent ears in infants to elude the need for surgical correction.

Otoplasty is usually performed under general anesthesia in children and with intravenous sedation in adults. The procedure involves an incision carefully hidden behind the ear; it can rarely be seen. Cartilage molding is done via otobrading, and suture reshaping/fixation to decrease ear projection. The ear is meticulously sculpted in an effort to have it look completely natural. (Figure 1b and 1d) A protective dressing is used continuously for one week followed by the use of a protective band at night for several more weeks. This technique has a low incidence of complications. Recurrences are rare, while patient and family satisfaction tends to be high.

Nose

The pediatric dictum that "children are not little adults" applies to nasal surgery. There are restraints imposed by immaturity of tissue as well as normal nasal growth. The size and shape of the nose changes dramatically from infancy to adulthood. Although there is no definitive consensus, many feel the nose should remain undisturbed until the postpubertal growth spurt.[8] By this time, the adolescent patient has a more developed personality. This development is necessary to address the complex concerns and issues necessary for rhinoplasty. Hence, rhinoplasty can generally be offered to an adolescent at approximately 16 years of age.

Rhinoplasty is performed to address the concerns of the patient and to achieve a natural appearing nose that artistically matches other features of the face. The nasal recontouring should enhance the artistic match of the nose to the face without obliterating ethnic and individual features. Thus, the operative plan is defined by these goals. The nasal skin thickness is an important variable because a thick skin sleeve limits what can be accomplished surgically.[13] The bony framework of the nose frequently needs to be altered in order to achieve the most pleasant shape and function. Techniques have been developed to perform this in a safe, reliable and reproducible way.[14,15] Overall, there should be a harmonious balance with surrounding structures including the cheeks, forehead and chin.

A nasal splint is worn for one week. Nasal packing is rarely necessary for rhinoplasty alone. Nasal airway obstruction can be addressed simultaneously during rhinoplasty when indicated. And we have found that both the septum and turbinates may require treatment in order to achieve normal function.

Chin

Aesthetic beauty of the lower face is dependent on the shape and position of the chin.[9] The posture and function of the lips also is influenced by chin position.[1] Overall, facial balance is determined predominately by the relationship of the nose and chin. [Figure 2a]. Genioplasty can be done to address a variety of mandibular positions. Key aspects to chin dysmorphology are anterior-posterior projection, inadequate or excessive vertical height, and depth of the labiomental crease.[10] Not only are these aesthetic concerns, but also functional considerations relating to malocclusion and mastication can be associated.[11]

Children and adolescents undergo rapid reorganization of the self-image after plastic surgery with subsequent positive changes in behavior and interpersonal interactions.

Timing mandibular surgery is variable. Genioplasty is usually considered after eruption of adult dentition as well as completion of craniofacial growth in adolescence. Preoperative photographs and cephalometric roentgenograms are part of surgical planning.[1] Numerous surgical approaches are available to the surgeon based on the patient's individual characteristics. These include alloplastic augmentation, sliding genioplasty, interposition alloplastic/autoplastic materials after horizontal osteotomy, and vertical shortening horizontal osteotomy. The field has continued to advance with the advent of plating systems to stabilize the bone in its new position after the osteotomy. [Figure 3B] Despite these excellent techniques, there is no substitute to accurate diagnosis of problems and careful planning.[12] Overall, genioplasty serves as a unique opportunity to improve facial aesthetics in select patients.[10]

Computer imaging can be used to alter photographs preoperatively and show the patient an image of how they might look postoperatively. The capabilities of computers and surgeons, though, do not mesh. Although the computer operator can easily make facial changes show up clearly on the screen, the surgeon may not be able to make those changes. Because computer-imaging may encourage false expectations, We avoid it.

Summary

Facial aesthetics can be enhanced by otoplasty, rhinoplasty and genioplasty. Excellent outcomes can be obtained given appropriate timing, patient selection, preoperative planning, and artistic sculpting of the region with the appropriate surgical technique. Choosing a patient with mature psychological, developmental, and anatomic features that are amenable to treatment in the pediatric population can be challenging, yet rewarding.

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Patrick K. Sullivan, MD, is Associate Professor, Plastic Surgery, Brown Medical School.

David P. Singer, MD, is a resident in the Department of Plastic Surgery, Brown Medical School.

Correspondence:
Patrick K. Sullivan, MD
235 Plain St., Suite 502
Providence, RI 02905
phone: (401) 831-8300
fax: (401) 831-8304
e-mail: cosmetic@drsullivan.com

Early Evaluation and Management of Craniofacial Dysmorphology

Patrick K. Sullivan, MD, and Chad D. Tattini, MD

Great outcomes and early detection go hand in hand. This is especially true in definitive treatment of head shape abnormalities, which relies heavily on the timing of the diagnosis. Optimal conservative measures and operative techniques vary according to the age of the patient, the presence or absence of functional impairments, and the extent of the abnormality. For these reasons early diagnosis is so important. To optimally care for this population, all clinicians should know the pathogenesis and the key features of the physical examination. Prompt diagnosis of the abnormality will ultimately improve treatment and increase the chances for a successful outcome for the patient.

Virchow in the 1850s first devised his law describing the natural growth of cranial sutures: the normal growth pattern of the calvarium is perpendicular to the patent suture; however, in synostoses the growth pattern is parallel to the affected suture.[1] Consequently, a skull can form into many different shapes, depending on the pattern of suture pathology.

The crucial period of craniofacial organogenesis takes place in the first 12 weeks of gestation. During this time the majority of congenital craniofacial anomalies are established. The cranial bones undergo ossification primarily through the intramembranous pathway.[2] Bone growth occurs primarily through the displacement of the calvarial bones by the enlarging underlying brain, which then stimulates deposition and remodeling of new bone in the spaces created.[1] At 6 months, the brain volume and cranial capacity is 50% of the adult brain volume; by the end of the first year, the brain volume has nearly quadrupled. Consequently, normal calvarial development relies heavily on this rapidly expanding brain.[3] This is absolutely key in the treatment of both plagiocephaly due to deformation and also for true craniostenosis.

The pathogenesis of craniosynostosis is essentially divided into two main schools of thought. Virchow explained that the calvarial suture is the primary abnormality, with the cranial base being the secondary deformity. Conversely, Moss proclaimed that the cranial base is the primary deformity, resulting in secondary cranial deformities. Moss supports this statement through his concept of the "functional matrix" which emphasizes the importance of the dura in suture patency. The dura allows substances to diffuse across a semipermeable membrane, thus keeping the suture patent. In addition, the dura has been shown to house osteoprogenitor cells.[1] It is thought that cranial base abnormalities are transduced to the calvarium and surrounding soft tissue via the dura.

A detailed history focusing on the family and prenatal history is essential to an early and effective diagnosis. Evaluation of the patient then proceeds with an intense analysis of the morphological abnormality. In addition, any functional impairment — (i.e. hydrocephalus, mental retardation, increased **intra-cranial pressure** (**ICP**), visual problems, or developmental delay) — must be evaluated. The physical examination also is helpful in classifying the problem into a syndrome, if one is present. Finally, plain skull radiographs may be used as a supplement, always being mindful of false negatives. A skull series is usually easy to obtain and often gives helpful information in a child with a head shape anomaly. It is usually worthwhile to delay further imaging until the craniofacial team has had the opportunity to evaluate the patient. Frequently, a specific type of exam may be needed, with a certain thickness of each CT slice in order to obtain the appropriate two-dimensional or three-dimensional [Figure 1] study. CT scans usually require sedation and increase radiation. Furthermore, three-dimensional studies may give false positive results when used diagnostically.[6] We feel CT scans can often be avoided if the suspicion for synostosis is low.

When evaluating the upper face and the cranium, there are helpful guidelines in the physical examination to help differentiate craniosynostoseis from deformational head shape abnormalities. The differentiation can frequently be made based on the physical exam alone. Deformational or positional plagiocephaly is not true suture synostosis, but rather a condition likely as a result of external compression forces on the calvarium in utero, at birth, or postnatally. Deformational plagiocephaly is much more common: it is estimated to be present in one in ten otherwise healthy infants. By contrast, the incidence of craniosynostosis in the general population is estimated at only 1 in 1000 to 2000 live births.[4]

We use a series of steps to differentiate between deformational plagiocephaly and plagiocephaly from synostosis. First, looking from above,

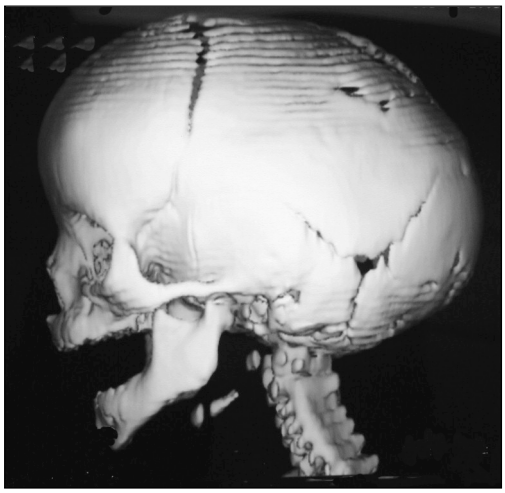

Figure 1 Three Dimensional study of a child with sagittal synostosis.

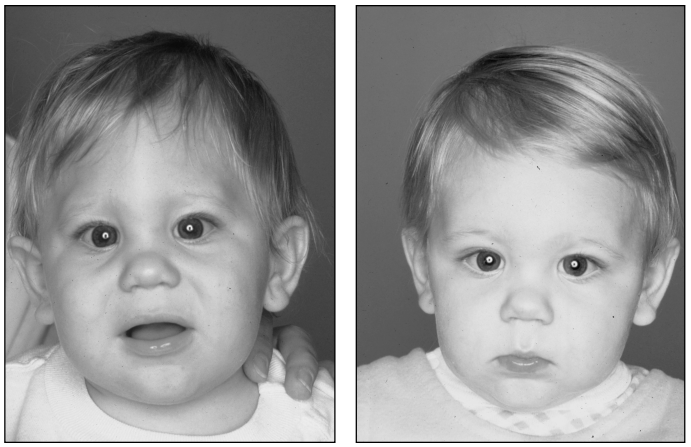

Figure 2A. A Child with Metopic Craniostenosis producing trigonocephaly with a V-shaped appearance of the skull. Figure 2B. After cranio-orbital reshaping of the forehead, superior and lateral orbits, anterior and lateral calvarium.

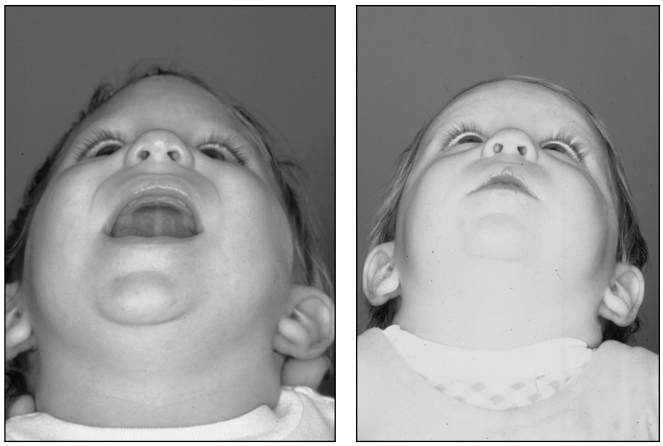

Figure 3A. The forehead has a central ridge with temporal flattening with the widest point being in the occipital region. Figure 3B. The stenosed suture and the forehead ridge have been removed. The temporal area has been significantly widened.

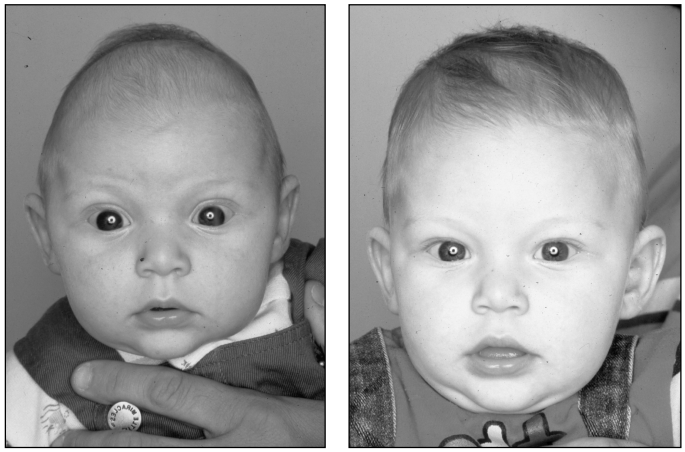

Figure 4A. Craniosynostosis with sagittal suture involvement has created a transverse narrowing of the calvarium. The AP calvarial distance has been significantly increased. Figure 4B. The transverse width has been increased.

Figure 5A. There is a saddle shape present to the calvarium. Figure 5B. The saddle shape has been removed.

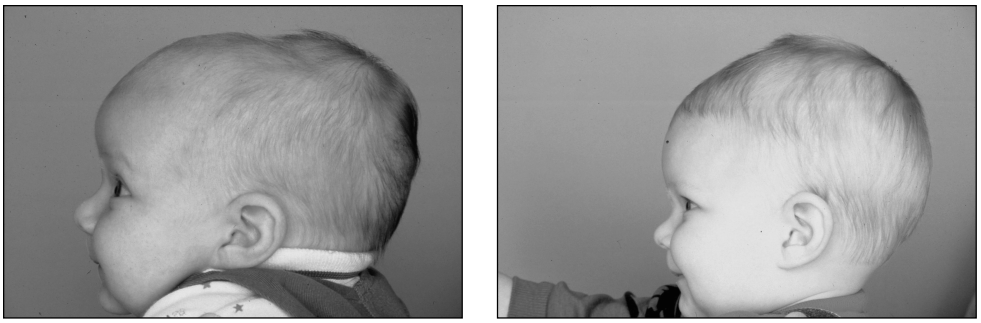

Figure 6A. There is an increased anterior-posterior distance. Figure 6B. The anterior-posterior distance has been decreased through craniofacial reshaping.

Table I

Differentiating Deformational Plagiocephaly from Plagiocephaly due to Craniostenosis

	Deformational	Craniostenosis
Ipsilateral:		
Forehead	flat	flat
Superior orbital rim	down	up
Palpebral fissure	slit-like	round
Malar	post.	ant.
Ear	post.-inf.	ant.-sup.
Nasal root:	midline	towards ipsilat.
Chin point:	towards ipsilat	towards contralat.

the side of the flattened forehead is identified. Second, on the side of the flattening, we evaluate the superior orbital rim, which may be superiorly positioned in synostosis. The root of the nose is specifically examined, as it usually is midline in deformational plagiocephaly, but deviated toward the side in unilateral coronal synostosis. We then examine the palpebral fissure, which has a slit-like appearance with deformational plagiocephaly and a round shape with synostosis. Finally, we inspect the ear as it is posterior and inferior on the ipsilateral side in deformational plagiocephaly and is anterior and superior in true synostoses.[5] (Table 1).

Deformational plagiocephaly usually improves with conservative measures and rarely needs surgical correction. It is recommended that the infant sleep prone or on the opposite side of the flattening. Helmet modeling may be necessary: the infant wears a helmet 15 to 23 hours per day (hrs/day) with follow-up every 2 to 3 weeks for adjusting.[1] This re-emphasizes the importance of early diagnosis since the efficacy of helmet modeling is drastically reduced after about 9 months of age when the exponential cranial growth is tapering.

The correct diagnosis of the head shape abnormality is essential since the treatment for each deformity is so different. Timing is absolutely key. When the child does need surgery, we may find it an advantage to operate within the first three months after birth. This depends on the diagnosis, the severity of the deformity, the number of cranial sutures involved, and the presence of increased intracranial pressure. One of the advantages of early surgery is that the bone is still malleable, and therefore easily shaped into the desired form. Another advantage is the rapid reossification rate of the infant cranium. Bony defects in the infant are usually completely ossified as long as surgery is performed during the first year of life. Also, the outward push of the growing brain is a key element supporting the reconstruction and is an advantage of providing treatment early during the first year of life. Beneficial effects also are seen on adjacent structures in the face when stenosed areas are released early in life. Finally, the psychological trauma is less on the patient and parents when the surgery is performed early in life.[2] [Figures 2- 6.]

The ultimate goals of craniofacial surgery … are twofold. First is the prevention of functional impairments. Second is to produce an aesthetically pleasing result that brings the child into the range of normal.

The ultimate goals of craniofacial surgery in the management of craniosynostosis are twofold. First is the prevention of functional impairments. Second is to produce an aesthetically pleasing result that brings the child into the range of normal. Advances in the management of these children and their families have come through a team approach with the primary-care specialists, pediatric services at our children's hospital, pediatric anesthesiologists, and an experienced craniofacial team. This team approach provides the most efficient and effective means of evaluating a challenging cranial abnormality early in the patient's life. Subsequently, the coordination of appropriate care tailored to the individual needs of each child facilitates a great outcome.

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Patrick K. Sullivan, MD, is Associate Professor, Department of Plastic Surgery, Brown Medical School.

Chad D. Tattini, MD, is a resident in the Department of Plastic Surgery, Brown Medical School.

Correspondence:
Patrick K. Sullivan, MD
235 Plain St., Suite 502
Providence, RI 02905
phone: (401) 831-8300
fax: (401) 831-8304
e-mail: cosmetic@drsullivan.com

Management of Pediatric Head Shape Abnormalities: Craniosynostosis and Positional Posterior Plagiocephaly

Charles E. Weaver, MD, PhD, and John A. Duncan, MD, PhD

Evaluation of head shape abnormalities are common reasons for referral to a pediatric craniofacial team of surgeons which include plastic surgeons and neurosurgeons. **Craniosynostosis (CS)**, the premature closure of one or more of the cranial sutures that can lead to abnormal skull shape and dysmorphic facial features, is a relatively rare condition present in only 10-16:10,000 live births in the United States.[1] The majority of referrals for head shape abnormalities are for unilateral flattening of the occipital region (posterior plagiocephaly). On the other hand premature fusion of the lambdoidal suture causing posterior plagiocephaly is the rarest form of single suture CS. However, coincident with the 1992 recommendation by the American Academy of Pediatrics favoring the supine position for infant sleep to avoid **sudden infant death syndrome (SIDS)** an increase in the incidence of posterior plagiocephaly was noted.[2] Initially many of these infants underwent surgical corrections under the diagnosis of CS. Recently it has become clear that the majority of posterior plagiocephaly is secondary to positional flattening and can be managed conservatively, using either positional measures to avoid continued focused pressure or corrective helmets or bands. The differentiation between true lambdoid CS and positional flattening remains a controversial aspect of pediatric craniofacial surgery. We will review the current understanding of CS and present our algorithm for the diagnosis and management of head shape abnormalities with special attention to positional posterior plagiocephaly.

Classification/Terms

Primary CS is defined as idiopathic isolated premature fusion of one or multiple cranial sutures resulting in characteristic head shape changes. Secondary CS is due to a known underlying condition that leads to premature fusion of one or multiple sutures. Simple CS denotes single suture involvement, whereas compound CS refers to multiple suture involvement. Syndromic CS denotes certain named syndromes with CS, usually compound, in association with other abnormalities. Nonsyndromic CS is isolated single suture involvement that may be familial. Many descriptive terms are used to refer to the characteristic head shapes produced by the fusion of one or multiple sutures. For example: * Scaphocephaly and dolichocephaly refer to the narrow elongated head with frontal and occipital projections caused by sagittal suture synostosis. * Clinocephaly is used to describe severe scaphocephaly associated with indentation of the vertex.

Prenatal exposure to smoking, cocaine, high altitude, and thyroid medication are risk factors for CS…

* Trigonocephaly is a triangular shaped head associated with hypotelorism and is caused by metopic suture synostosis.
* Anterior plagiocephaly caused by unilateral coronal suture synostosis leads to ipsilateral flattening and contractual bossing of the forehead, ipsilateral elevation of the orbit (harlequin eye deformity), and ipsilateral deviation of the root of the nose causing the tip of the nose to appear to deviate contralaterally.
* Posterior plagiocephaly describes the occipital flattening that can be caused by unilateral lambdoid synostosis or by positional molding.
* Brachycephaly is the broad skull caused by either bilateral coronal or lambdoid synostosis.
* Kleeblattschadel, meaning cloverleaf skull is caused by fusion of the coronal, lambdoid, and metopic sutures.
* Acrocephaly, oxycephaly, turricephaly, and pyrgocephaly all refer to patterns of multiple fused sutures, usually syndromic, leading to severe restriction of cranial volume and are frequently associated with increased intracranial pressure.

Pathogenesis

In contrast to the bones of the skull base which, like long bones, grow by the process of endochondral bone growth, the bones of the calvarium form by a process called intramembranous bone growth.[3] Cells from the neural crest form the ectomeninx, the dural anlage. Ossification centers form (13wks) within the ectomeninx and bone growth progresses outward. At 18wks the mineralizing bone plates meet and sutures are formed along the various junctions. Calvarial growth from this point is induced by expansion of the growing brain,[1] which depends on an equilibrium at the sutures. Here, two plates of bone, comprised of undifferentiated rapidly dividing osteogenic stem cells are separated by a narrow space. A proportion of stem cells are recruited to differentiate into osteoblasts and form new bone at the growing bone margin while sufficient undifferentiated cells must remain to maintain the structure of the suture and prevent fusion. In addition, remodeling occurs by a process of endocranial resorption and ectocranial deposition.[4] The majority of growth in the human skull occurs before the age of 2, with a doubling in size in the first 6 months and doubling again by 2 years.[5] The sutures subsequently fuse

leaving behind immobile suture joints. Detailed studies of single suture CS have led to a better understanding of the compensatory changes in cranial shape that develop. Virchow, who initially described the characteristic deformities associated with premature closure of specific sutures, observed that growth continues perpendicular to the plane of the fused suture.[3] These observations have been modified to include the notion that the majority of compensatory growth occurs at adjacent open sutures.[6]

The cause of nonsyndromic CS is not completely understood and is likely the result of many pathologic processes. Mechanical causes such as intrauterine constraint due to multiple pregnancies, oligohydramnios, or lodging of the fetal head in the pelvis have been suggested to cause some cases.[1,7] In a mouse model of intrauterine constraint 88% of pups are born with CS.[8] It is possible that mechanical constraint may lead to dysfunction of the suture, leading to premature fusion. Nonmechanical factors may also play an important role in the development of CS. The regulation of suture formation and the timing of closure is a complex process involving the bone itself as well as soluble signals from the underlying dura.[9] The molecular framework for this complex signaling pathway is beginning to be understood. Mutations in **fibroblast growth factor receptors** (**FGFR1-3**) and transcription factors (MSX2, TWIST) have been identified in patients with both syndromic and nonsyndromic craniosynostosis.[10,12]

…children in whom the initial diagnosis is unclear will declare as true lambdoid fusions when they fail to respond to helmet therapy.

Diagnosis

The diagnosis of CS is primarily based on clinical examination and confirmed by radiographic studies. Abnormal head shape in CS is usually present at birth and progresses with time. However, some infants born with a normal head shape develop a progressive deformity. Children with persistent or progressive deformity at 2 to 3 months of age should be referred to a craniofacial specialist to be evaluated for craniosynostosis. Children with disorders known to cause CS should be followed expectantly because of their increased risk.

The initial evaluation should include the prenatal, delivery, postnatal and family history. Prenatal exposure to smoking, cocaine, high altitude, and thyroid medication are risk factors for CS and should be noted.[13-15] Any history of intrauterine constraint (multiparity, oligohydramnios, and amniotic bands) and the use of forceps or vacuum assistance should be identified. A family history of CS, abnormal head shape, or neurologic disorders should be sought. The time the abnormality was first identified and whether it has progressively worsened or improved with time should be noted. Of particular importance is the identification of any head position preference or condition that cause it (torticollis or strabismus). Any evidence of increased intracranial pressure such as episodes of lethargy, disconjugate gaze, papilledema, developmental delay, and on plain x-rays or CT studies scalloping of the inner surface of the calvaria ("beaten copper"), dilated CSF spaces or transependymal CSF flow should be identified. On physical examination the areas of sutures should be palpated and any ridging noted. The state of the fontanels should be assessed. Any dysmorphic facial features should be noted, and the head circumference recorded. Plain skull x-rays should be obtained to confirm the fused suture(s) and demonstrate the characteristic shape changes associated with each specific suture closure. A minimum of four views of the skull should be obtained to adequately view all sutures. In cases where plain film studies are equivocal spiral CT with 3D reconstructions can be useful in confirming the diagnosis of a fused suture. CT studies are more sensitive than plain x-rays in detailing suture anatomy.[1] In addition, CT allows for evaluation of the brain apart from bone and may identify underlying abnormalities associated with synostosis. 3D models can be extremely useful in the preoperative planning when surgical repair is to be undertaken. Nuclear bone scans, although used in the past to demonstrate decreased activity at fused sutures, is currently not used. Ultrasound, although reported to differentiate between open and fused sutures[16] has not been widely utilized. MRI adds little to the workup.

The diagnosis of posterior plagio-

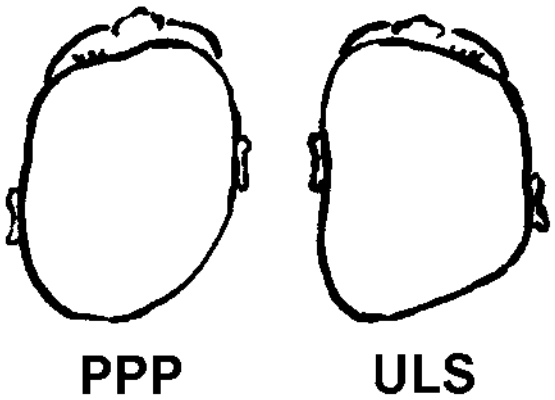

Figure 1. Head shape when viewed from above in posterior positional plagiocephally vs unilateral
lambdoid synostosis.

Table 1 Differentiation between ULS and PPP.

Feature	Posterior Positional Plagiocephaly	Unilateral Lambdoid Synostosis
Unilateral occipital flattening	Present	Present
Frontal bossing	Ipsilateral	Contralateral
Contralateral posterior bossing	Parietal	Occipital
Ear displacement	Anterior	Posterior
Displacement of Petrous bone	Anterior	Posterior
Rotation of foramen magnum	Absent	Present
Ipsilateral lambdoid ridging	Absent	Present
Lambdoid suture	Patent	Fused
Overall Shape (Fig. 1)	Parallelogram	Rhomboid

Adapted from Park and Shenandoah (2000).

cephaly has been a point of controversy during the past 10 years. It is now widely accepted that posterior plagiocephaly can be caused by either positional molding from persistent head position or true premature fusion of the lambdoid suture. The concept of positional deformation of the posterior skull is not new and was recognized long before the 1992 recommendation by the American Academy of Pediatrics that infants be placed supine for sleep.[17,18] The change in parent and subsequent positioning behavior preceeded the nearly six fold increase in the prevalence of posterior plagiocephaly observed from 1992 to 1994.[19] No craniofacial specialist would suggest abandoning the recommendation for supine infant sleep, given the significant protection this affords infants against sudden infant death. Rather parents and pediatricians should be aware that positional flattening can occur. The distinction between posterior positional plagiocephaly and unilateral lambdoid synostosis can most often be made on the basis of careful physical examination. Table 1 details the characteristic findings in each. Of note, the overall head shape in unilateral lambdoid synostosis is rhomboid in contrast to the parallelogram appearance in posterior positional plagiocephaly (Figure 1).

Management

General principles

The negative psychological impact of head shape abnormality and dysmorphic facial features from CS are difficult to quantify. One study of children with untreated CS suggests there is a negative impact on both the affected child as well as the family.[20] The morbidity associated with elevated **intracranial pressure (ICP)** is more obvious. Increased ICP can lead to papilladema, optic atrophy and irreversible visual loss. Although the incidence of increased ICP in single suture CS is low it is not insignificant. Patients with minimal forms of CS have been shown to present with clinically significant increases in ICP.[21] Increased ICP is more common and severe in syndromic CS and when multiple sutures are involved.

Whether there are restrictions to brain growth and development with CS of single sutures is not clearly known but SPECT studies have demonstrated abnormal perfusion of the brain under fused sutures that returns to normal following surgical repair.[22] The functional consequences of these perfusion changes are not known.

Surgery

It is generally accepted that surgery for craniosynostosis should be performed early, preferably within the first year of life, to minimize the progression of deformity and the risk of developing elevated ICP as well as to maximize the time for normal development of the face and skull.[17, 23-25] Early surgery has been demonstrated to lead to better morphologic results without a significant increase in morbidity or mortality.[25] The success of early surgery for CS has been brought about largely by the use of a team approach combining neurosurgeons, plastic surgeons and pediatric anesthesiologists. Whenever possible a zigzag type "stealth" incision is used to reduce the visibility of the scar.[26] The most significant risk to neonates undergoing craniofacial reconstructions is that of major blood loss, which can be life threatening. For this reason meticulous surgical technique and close monitoring of hemodynamic status are critical to ensuring favorable outcomes. We strive to minimize blood loss by infiltration of the scalp with epinephrine before the skin incision is made. Skin clips are used to further minimize scalp bleeding. The periosteum is left in place where possible to minimize bone bleeding. Injury to the venous sinuses is avoided and any bleeding from the sinuses is controlled immediately. An arterial line and a minimum of two peripheral intravenous lines are placed prior to surgery for continuous arterial pressure monitoring, serial assessment of hemoglobin concentration, and iv access, should transfusion of blood products be required. In addition, body temperature is rigorously maintained in the normothermic range to prevent disturbances in coagulation. Postoperatively all patients are maintained in an intensive care unit setting for a minimum of 24 hours for hemodynamic monitoring, frequent neurologic assessments and pain control. Intravenous hydration with isotonic saline to avoid hyponatremia is continued until infants are taking fluids by mouth. Antibiotics are started preoperatively and continued 24 hrs after subgalial drain removal.

Posterior Positional plagiocephaly and Unilateral Lambdoid synostosis

In any pediatric office today, occipital flattening is the most frequent head shape abnormality seen. The vast majority of these are positional in nature. In our pediatric neurosurgical clinic posterior positional plagiocephaly accounts for 20 times the number of all CS seen and 400 times the number of unilateral lambdoid synostosis. As discussed, the diagnosis is made most frequently on a clinical basis. If the in-

fant is young (2-3 months) and the positional deformity is mild a trial of positional therapy can be attempted. The parents or caretakers are counseled on positional measures to limit continued focal pressure. If torticollis is present physical therapy should be initiated. In infants with significant deformity or those who have failed to improve with a trial of positional therapy, we recommend the use of a moulding helmet. The helmet is designed to apply gentle pressure to prominent areas while allowing flattened areas to expand leading to a more symmetric configuration. Helmets are most effective in children during the first year of life as they take advantage of the rapid increase in head size during this period. The device is to be worn 23 hrs a day, allowing for bathing and to examine for areas of excessive pressure. Families work closely with an orthotist to ensure a good fit and are followed in our office every 2-3 months to assess progress. Head circumferences are followed at each visit. Head growth proceeds unhindered during the use of the helmet as has been documented with the dynamic orthotic cranioplasty band.[27] Improvement is usually evident after only 2-3 months and most children require between 4-6 months of helmet use for an excellent cosmetic result. The success rates for positional and helmet therapy has been reported to be between 51% and 89%[28-30] with one study suggesting better and faster results with helmet therapy when compared to positional therapy.[31] If suspicion of lambdoid synostosis is raised by the clinical evaluation, imaging studies including plain x-rays and CT scan with 3D reconstructions are obtained. If true lambdoid synostosis is confirmed surgical correction is indicated as these children will not respond to conservative therapies. If imaging studies are not definitive, we favor a trial period in a moulding helmet. In our experience, children in whom the initial diagnosis is unclear will declare as true lambdoid fusions when they fail to respond to helmet therapy. In our practice this is often seen at either 12 - 15 months of age or after 5 -6 months of helmet treatment. This treatment algorithm allows for selection of children with true lambdoid fusion and results in a very good, if not excellent, reconstructive result and as importantly, avoids unnecessary surgery and all its associated risks. In contrast to other forms of craniosynostosis, in which reconstruction is favored between 3 to 9 months of age, treating physicians and families should follow a different, more conservative course in the treatment of children with of children with posterior plagiocephaly.

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Charles E. Weaver, MD, PhD, is a resident in neurosurgery, Rhode Island Hospital.

John A. Duncan, MD, PhD, is chief of neurosurgery, Rhode Island Hospital and Hasbro Children's Hospital, and Assistant Professor of Neurosurgery, Brown Medical School.

Correspondence:
Charles E. Weaver, MD, PhD
Rhode Island Hospital
593 Eddy St.
Providence, RI 02903
phone: (401) 444-8716
fax: (401) 444-3422
e-mail: Charles_Weaver_MD@brown.edu

Hearing Loss in Children with Craniofacial Anomalies

Brian E. Duff, MD

Children born with craniofacial anomalies are at an increased risk of hearing loss. This association is often the result of abnormal development along the auditory pathway and can occur anywhere from the pinna to the central nervous system. If problems arise in the outer ear or middle ear, patients will manifest a conductive or mechanical hearing loss. A sensorineural or neural hearing loss occurs with abnormalities of the cochlea, auditory nerve or central nervous system. Patients with a mixed hearing loss have both a conductive and sensorineural component. For the purpose of this discussion we will examine the etiology of these patterns of hearing loss.

Conductive Hearing Loss

The pinna develops from the upper end of the first and second branchial arches and from the upper end of the first branchial cleft.[1] This development is complete by the fourth gestational month. During the second month a solid core of epithelium migrates inward from the rudimentary pinna toward the first branchial pouch. This core of epithelium is the precursor of the external auditory canal. The first branchial pouch grows outward to form the tympanic cavity or middle ear cleft. The plaque of tissue formed where this meatal plug and the first branchial pouch meet ultimately becomes the tympanic membrane. At the same time the malleus and incus begin taking shape from the upper end of the first branchial arch. The stapes forms from the upper end of the 2nd branchial arch and all the ossicles attain their final shape by the end of the fourth month. During the sixth month of gestation the external canal begins to hollow out and this process is completed by the end of the seventh month. The extent of any developmental abnormality depends on the timing of the insult. The earlier the disruption, the more severe the deformities of the pinna, external canal and middle ear.

Atresia of the ear canal is often associated with abnormalities in the size and/or shape of the pinna (microtia). These ear malformations may occur in isolation or in association with other craniofacial dysplasias. The reported incidence of aural atresia is 1 in 10,000 to 1 in 20,000 births.[2] Genetic transmission occurs in many of the syndromes that include aural atresia (e.g. Crouzon's syndrome and Treacher-Collins syndrome). Isolated cases of atresia usually represent a sporadic mutation rather than genetic inheritance.

Patients with congenital aural atresia are classified on the basis of auricular development and external canal/middle ear development. Deformity of the auricle is straightforward and is divided into three grades. Grade I microtia represents a minor malformation, with the auricle being smaller than normal but with all parts discernible. In grade II microtia, the auricle is represented by a curving or vertical ridge of tissue. In grade III microtia, any resemblance to an auricle is lost, and only a small rudimentary soft tissue structure is present.[2]

Isolated deformities of the auricle are not associated with a significant conductive hearing loss and reconstruction is undertaken for the purpose of cosmesis. Surgery is usually performed just before the child enters school when most of the growth of the auricle has occurred. In cases of severe deformity, osteointegrated implants may permit the use of an auricular prosthesis rather than requiring the patient to undergo a multiple stage surgical reconstruction.

Atresia of the external canal / middle ear deformity is divided into minor and major malformations. Absence of the external ear canal and tympanic membrane characterize the major malformation group. Also included in this group are cases of severe canal stenosis. The size of the middle ear space is reduced and the malleus and incus are frequently deformed and fused. In severe cases, the middle ear space is hypoplastic and the ossicles are absent. In the minor malformation group the external canal is often small but patent while the middle ear space is normal in size. A conductive loss exists due to deformity of one or more ossicles and/or fixation of the ossicular chain.[2]

Management of the conductive hearing loss associated with canal atresia depends on whether the deformity is unilateral (70%) or bilateral. In cases of unilateral atresia the first priority is to determine the status of the contralateral ear. If audiometric testing confirms normal hearing in the normal appearing ear, the parents can be reassured that normal speech and language development is probable. The child should then be evaluated at regular intervals to determine whether intervention (i.e. speech therapy and/or amplification) is required. In infants with bilateral atresia, early amplification is essential. The initial medical and audiologic evaluations should be completed as quickly as possible and the child fitted with an appropriate hearing aid.[3]

Approximately one third of patients with aural atresia are not candidates for surgical correction. Important in the selection process are the physical examination, audiometric assessment and CT appearance of the temporal bone. In most cases it is the radiographic findings of the middle ear that determines candidacy. In general, radiographic evaluation is delayed until around 4 years of age to permit growth of the temporal bone and mastoid air cell development to occur. A temporal bone CT should be done earlier if there are suggestions that otitis media in an atretic or stenotic ear may have caused complications such as meningitis, facial paralysis or suppurative labyrinthitis. If microtia is present then reconstruction of the external canal can be considered after auricular reconstruction has been completed. Atresia surgery is usually performed when the child is 5 to 6 years of age. By this time, accurate audiometric tests have been obtained, pneumatization of the temporal bone is well advanced and children are capable of cooperating with postoperative care. To eliminate the handicap of unilateral hearing loss, a threshold of 25dB or better must be achieved. Of ap-

propriately selected patients, 70% will enjoy this degree of postoperative hearing recovery.

Cleft palate

Recurrent middle ear infections and associated conductive hearing loss occur in 85-95% of children with a cleft palate deformity. Bluestone et al[4] have shown that **Eustachian tube (ET)** dysfunction in cleft patients occurs because tubal dilator muscles, which have palatal insertions, are less effective. These patients cannot completely equilibrate air pressure between the middle ear and the nasopharynx. In the underventilated middle ear, gas is resorbed and the resultant negative pressure creates the diffusion gradient for serous effusion to occur.

In addition to its ventilatory role, the ET also prevents reflux of nasopharyngeal secretion into the middle ear.[5] In children with a cleft palate oral secretions and feedings freely enter the nasopharynx. Thus the presence of a palatal cleft may result in a nutrient-rich culture medium within the middle ear and the means by which it becomes inoculated.

The effects of binaural chronic middle ear effusions on speech and language development have been extensively studied. Though there is still some debate, most of the research identifies problems with articulation, vocabulary, grammar, syntax, auditory memory / processing and auditory-visual integration.[6] The indications for surgery should reflect these concerns for compounding the speech and language deficits for which cleft palate patients are prone.

Though the incidence of chronic otitis media decreases after palatoplasty, most children will meet conservative criteria for ventilating tube insertion both before and after palatal repair.[7] Recommendations for tube insertion and timing should be made on an individual basis with an emphasis on long-term surveillance.

Sensorineural Hearing Loss

Children with craniofacial anomalies are at an increased risk for sensorineural hearing loss. This loss may have a syndromic association (Apert's syndrome, Stickler's syndrome) or be a sporadic event. Development of the inner ear is well advanced by the end of the 8th week when the membranous labyrinth has assumed its characteristic convoluted shape. Gradual ossification of the otic capsule around the membranous labyrinth is essentially complete at birth. Maturation of hair cells and auditory neural development occurs during the late second and early third trimester so that the normal fetus may be able to hear 2.5 to 3 months before birth.[8]

Most inner ear malformations arise when formation of the membranous labyrinth is interrupted during the first trimester of pregnancy. Abnormalities in otic capsule structure and deficiencies in the cochlea appear to arise as secondary effects of the earlier error in development of the membranous labyrinth. Only 5-15% of congenitally deaf individuals demonstrate a bony abnormality on temporal bone CT.[9]

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There is a wide spectrum of severity of hearing disability among these patients. Although some individuals are deaf from birth, most maintain residual hearing into adulthood. Sudden hearing loss is frequent, either spontaneous or as a result of head trauma, even minor in nature. Vestibular symptoms, which are occasionally severe, are present in 20% of patients.[8]

High resolution temporal bone CT imaging is recommended for all children with an otherwise unexplained sensorineural hearing loss as soon as the loss is first recognized. When possible, axial and coronal 1.0mm images are obtained. Recognizing an inner ear malformation on CT scan may prove helpful as certain morphologic patterns have more favorable prognoses than others.

No medical or surgical therapy has been shown to prevent the progressive hearing loss associated with congenital malformation of the inner ear. Children with inner ear anomalies should avoid head trauma (contact sports) and rapid barometric changes (scuba diving or unpressurized aircraft) as these risk a sudden loss of hearing or even CSF leakage. For those children with radiographic risk factors, parents should be made aware of the risk of meningitis and instructed in recognizing its early signs. As most episodes of meningitis associated with inner ear malformation are pneumococcal, use of the pneumococcal vaccine is recommended. For the hearing impaired child, it is useful to establish a prognosis for future auditory function, as this serves as a guide for educational and rehabilitative efforts.

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Brian E. Duff, MD, is Director of Otology/Neurotology at Rhode Island Hospital/Hasbro Children's Hospital and Assistant Clinical Professor of Otolaryngology, Brown Medical School.

Correspondence:
Brian E. Duff, MD
100 Dudley St.
Providence, RI 02905
phone: (401) 274-2300
fax: (401) 272-1302
e-mail: BDuff@lifespan.org

Speech, Language, and Hearing Management of the Child with Cleft Palate

Paul W. Austin, MEd, CCC SLP/A

Management of the child with a cleft lip or palate necessitates a team approach. The problems that the child with cleft lip and/or palate faces require the skills of the plastic surgeon, dentist, orthodentist, otolaryngologist, pediatrician, geneticist, speech/language patholgist, audiologist. This section will deal with the role of the speech/language pathologist and audiologist on that team.

Often the resident in plastic surgery is asked, "Why repair a cleft palate?" The answer is, "To give the patient better speech." Speech is a major focus in the management of the child with cleft palate. [We are restricting our focus in this article to children with cleft palate since rarely do speech problems occur in children who have a cleft of the lip only.]

Speech is a broadly used term to describe multiple facets comprising a child's communicative ability. In order to assess the child with cleft palate it is necessary to evaluate the following: hearing, receptive language, expressive language, articulation, resonance, and voice.

Hearing

Hearing assessment is a vital part of the evaluation battery in children with cleft palate. Rhode Island was one of the pioneering states in mandating hearing screenings for newborns. This allows for early identification of hearing impairment in the child with cleft palate. Reports have suggested that 95-100% of infants born with cleft palate have fluid present in the middle ear space,[1] which is usually correlated with the presence of a conductive or middle ear hearing loss The early identification of hearing loss allows for timely and early treatment. This early identification also allows for a more comprehensive audiologic evaluation using more objective measures such as Brainstem Evoked Response Audiometry , Tympanometry, and Otoacoustic Emissions. These measures yield a more accurate profile of the child's hearing, while allowing for timely treatment by the otolaryngologist.

The impact of minimal hearing loss (20 decibels) on a child's speech and language development has been argued greatly in the literature. McWilliams[2] (1990) suggests that children with middle ear disease who demonstrate chronic conductive hearing loss should at the least have preferential seating in the classroom. If the hearing loss becomes significant it may interfere with both educational and social functioning.[2] If the loss extends into the moderate range (40-55dB) and is bilateral (and medical treatment has not been successful) use of hearing aids or other assisistive listening devices should be strongly considered.[2]

It is difficult to recommend a schedule of hearing assessment in the child with a cleft palate. If a hearing loss is identified as a newborn there are follow up protocols already established through the R.I. Hearing Assessment Project. Once treatment has been completed, the child should be monitored audiologically at 3-6 month intervals.

Receptive and Expressive Language

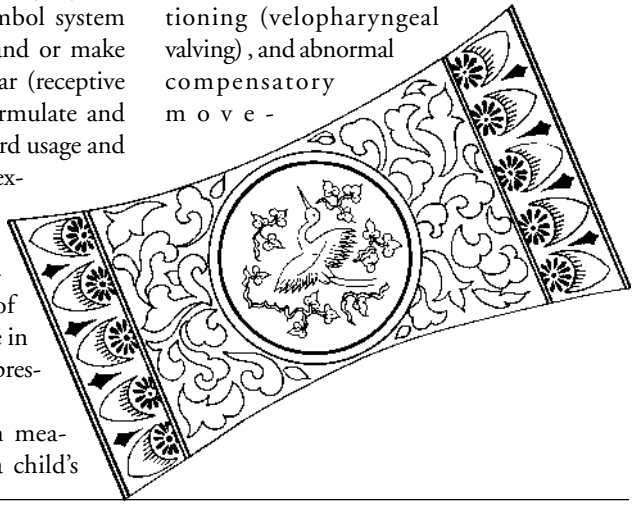
When we talk about language we are talking about the symbol system which we use to understand or make sense of the words we hear (receptive language) and then to formulate and speak with appropriate word usage and appropriate word order (expressive language.)[3] Part of the assessment of the child with cleft palate necessitates an evaluation of their language competence in both the receptive and expressive domains.

Objective evaluation measures exist which assess a child's competence in both the receptive and expressive language areas. Certainly it is important to educate parents about the language development milestones. Parents serve an important role as "evaluators" of their child's language competence and can advocate for early evaluation and intervention. Normally children by the age of 12 months begin using a single word; by 18 months are combining two words; and by two years are using two and three word phrases. If the child with cleft palate is found to have deficits in any one of these areas, speech and language therapy intervention can be put in place.

Speech

Speech refers to the motor component of our communication. In order for speech to develop in a normal fashion there must be intact function of the lips, jaw, tongue, teeth and palate. In our Cleft Palate-Craniofacial Center we like to see the child around the age of 18 months. This allows for an assessment of his receptive and expressive language and also to begin to make an inventory of his speech development.

The child with cleft palate can present with some unusual speech (articulation) patterns. This can be related to abnormal dental allignment, abnormal lip configuration, poor tongue mobility; poor soft palatal functioning (velopharyngeal valving) , and abnormal compensatory move-



ments.

Early identification of articulation deficits allows for initiation of appropriate speech therapy services with a strong emphasis on parent guidance. Parents can be taught strategies to carry treatment methods over into the home.

Early speech evaluation also allows for the detection of some of the abnormal compensatory articulation errors that are sometimes seen in the child with cleft palate. Some of these errors are pharyngeal frication (air flow is trapped at the back of the throat on sounds such as /s/ and /sh/; and glottal stop (air is trapped where the sound is produced or at the vocal folds.)

Resonance

Resonance, a difficult term to define, is a major factor to assess when evaluating the speech of a child with cleft palate. In the patient with a normal palate, the oral and nasal cavities are not coupled but remain separate except on the 3 nasal consonants of the English language ('m', 'n', and 'ng'.) When there is a coupling or joining of the oral and nasal cavities, speech is perceived as nasal or hypernasal. The presence of hyperenasal resonance is usually associated with lack of adequate function of the soft palate (velopharyngeal incompetence.)[2] The soft palate acts as a sphincter and prevents air from escaping into the nasal cavity except on the 3 previously mentioned sounds. This sphincteric action also prevents food or liquid from entering the nasal cavity. One of the important questions during the case history with younger children in a pre speech phase is the occurrence of any food or liquid "leaking" through the nose. This could be an early indication of velopharyngeal incompetence. Resonance evaluation of the child with cleft palate is important at a single word phase to determine whether hypernasality is present on specific isolated sounds.[4] It is also important to assess connected speech because hypernasality may become more evident at this level which places greater demands on the velopharyngeal valving mechanism. In our center we tape-record connected speech samples as one means of determining the need for surgical management to correct velopharyngeal incompetency. Other evaluation tools may include completion of fiberoptic nasopharyngoscopy and more recently flexible scope video nasendoscopy. Both procedures allow for viewing of the velopharyngeal mechanism in its dynamic state in children usually 5 years of age or over.[5]

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Voice

Children with cleft palates will sometimes develop a vocal hoarseness, usually a result of the presence of vocal cord nodules demonstrated on laryngoscopy.[2] This condition can result from vocal hyperfunction. The patient who presents with velpharyngeal imcompentency will often constrict the muscles of the throat as a means of trying to reduce the hypernasal resonance quality. Surgical management of the velopharyngeal incompetency and behavior therapy to reduce the vocal hyperfunction will usually result in a normal vocal and resonance quality.

Summary

The management of the patient with cleft palate requires a multi-disciplinary team approach. The end result, from a speech, language, and hearing perspective is to have a child enter school at age 5 with as near normal a speech profile as possible. This is due to an assessment that looks at receptive and expressive language, hearing, speech, and resonance. Early assessment and detection of problems then allows for timely and early intervention and essential parent guidance.

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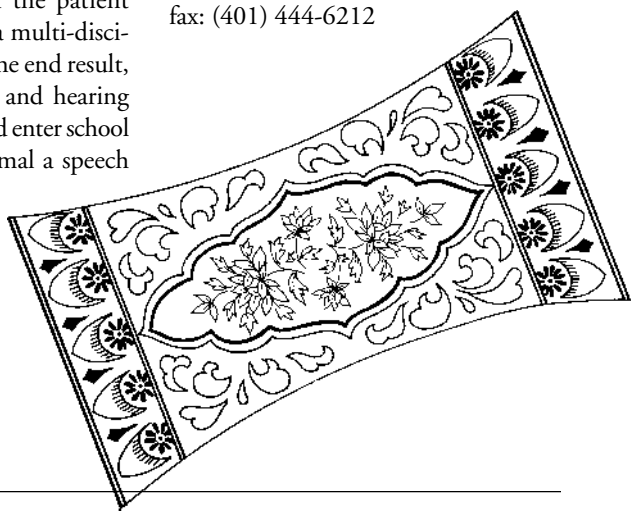
Paul W. Austin, MEd, CCC, is Speech Pathologist/.Audiologist at Rhode Island Hospital/Hasbro Children's Hospital, Department of Speech Language Pathology/Audiology; and Speech Pathology/ Audiology Consultant, Cleft Palate-Craniofacial Team, Rhode Island Hospital.

Correspondence:
Paul W. Austin, MEd, CCC SLP/A
Audiology/Speech Pathology
Rhode Island Hospital
593 Eddy St.
Providence, RI 02903
phone: (401) 444-4054
fax: (401) 444-6212



Airway Management in Children with Craniofacial Anomalies

Sharon E. Gibson, MD

Upper airway obstruction is common in children with **craniofacial anomalies (CFA)**; 60% of children with symmetric skeletal abnormalities of the maxilla, mandible, or both will require intervention for symptoms of upper airway obstruction.[1] Eighty percent of these will present within the first month of life.

A myriad of factors, involving both bony and soft tissue structures, contribute to upper airway obstruction. Maxillary and mandibular hypoplasia, macroglossia, glossoptosis, excessive pharyngeal soft tissues, poor neuromuscular tone, and poor handling of pharyngeal secretions are common in the CFA patient population. Congenital laryngeal and tracheal anomalies also occur with increased frequency. Many patients suffer a combination of factors with obstruction at multiple levels of the upper airway.

Upper airway obstruction may present acutely at birth, with symptoms of stridor, stertor, retractions and tachypnea. Increased work of breathing while feeding may result in feeding difficulties and failure to thrive. Infants with CFA are four times more likely to have feeding problems when symptoms of airway obstruction are present as when there are no airway issues.[1] Some children do well initially, but develop obstructive sleep apnea over time as growth of the pharyngeal soft tissues occurs within a constricted bony framework. Surveillance for symptoms of upper airway obstruction is therefore necessary throughout childhood.

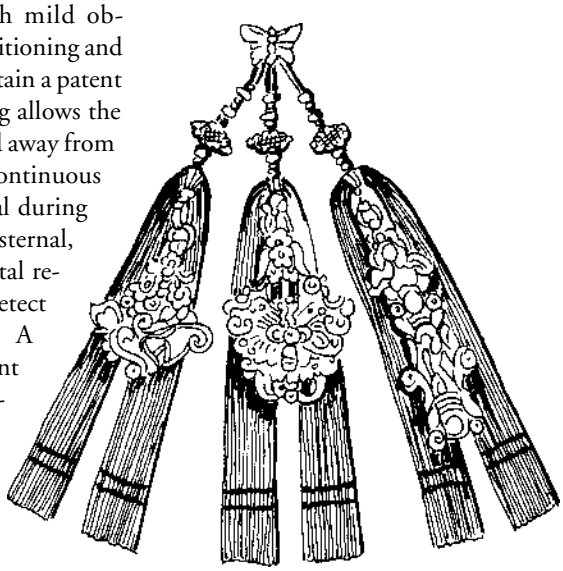
Airway Issues in the Cleft Palate Patient

An isolated cleft palate does not present with airway obstruction in infancy. Neonatal upper airway obstruction primarily concerns patients in whom cleft palate is a manifestation of an underlying syndrome. In the cleft palate syndromes, of which **Pierre Robin sequence (PRS)** is the classic example, abnormal development of the adjacent skeletal and soft tissues is responsible for the obstruction. In infants with PRS, the etiology of airway compromise is multifactorial and includes hypotonia of the pharyngeal soft tissues and hypoplasia of the jaw. Underdevelopment of the mandible during gestation results in a constricted oral cavity, which displaces the tongue superiorly and posteriorly. This abnormal tongue position in utero prevents closure of the palatal shelves, resulting in a wide cleft. At birth, the tongue may fall back sufficiently to cause glossoptosis, the apposition of the tongue base to the posterior pharyngeal wall and soft palate, obliterating the pharyngeal airway. Glossoptosis occurs in approximately 75% of children with PRS[1] and can result in severe, life-threatening respiratory distress.

Management of the airway is dictated by the degree of respiratory compromise. The major emphasis in treating infants with PRS has focused on (a) positioning; (b) pharyngeal stenting devices; (c) surgical procedures to draw the tongue forward; and (d) tracheostomy.

Many children with mild obstruction require only positioning and feeding strategies to maintain a patent airway. Prone positioning allows the tongue to fall forward and away from the posterior pharynx. Continuous pulse oximetry is essential during the initial trial, since substernal, suprasternal and intercostal retractions are difficult to detect in the prone infant. A nasogastric tube may stent the soft tissues of the pharyngeal airway adequately and allows concomitant gavage feedings to eliminate the increased work of breathing associated with oral feeds. Greater stenting is achieved by a nasopharyngeal tube (nasal trumpet). Nasopharyngeal airway devices are relatively tenuous, as slight migration of the tube tip inferiorly causes choking and emesis and superior displacement fails to maintain airway patency, and accidental dislodgement is common. Nasopharyngeal tubes are most suitable for short term inpatient use while other treatable causes of airway obstruction, such as gastroesophageal reflux disease, are sought, identified and treated. Via such strategies, the airway can be managed conservatively in up to 70% of newborns with PRS.[2,3]

Surgical intervention is warranted when apnea, stridor, retractions, and feeding difficulties persist despite conservative measures. The airway must initially be secured by endotracheal intubation which can be extremely challenging in PRS, since the small mandible restricts lifting of the tongue base during laryngoscopy, providing poor exposure of the larynx. The flexible fiberoptic bronchoscope and laryngeal mask airway have proven to be invaluable tools for achieving intubation in particularly difficult cases. Surgical procedures focusing on the



elimination of glossoptosis have been largely disappointing. The tongue-lip adhesion method of glossopexy does not provide adequate forward distraction of the obstructing tongue base and is not recommended for moderate or severe upper airway obstruction.[3,4] Hyomandibulopexy provides greater distraction of soft tissues but still fails to stabilize the airway in one third of patients so treated. Unfortunately, this procedure anchors the larynx more anteriorly, rendering the airway more difficult to intubate should the procedure fail and may interfere with mandibular growth. A subperiosteal release of the floor of mouth musculature from the mandible, performed through a submental incision, theoretically provides rotation of the tongue base[4] and has been successful in alleviating upper airway obstruction in select cases. Most infants with airway distress failing conservative therapy require surgical bypass of the pharyngeal obstruction by tracheostomy. A thorough endoscopic evaluation by direct laryngoscopy and bronchoscopy is performed at the time of surgery, to detect associated congenital airway anomalies. The presence of multiple airway abnormalities, such as laryngomalacia, subglottic stenosis or tracheomalacia, increases the chance that tracheostomy will be needed. Children with multisystem anomaly syndromes and concommitant neurologic disease are more likely to require tracheostomy (40%) as compared to those with PRS (8%).[1] Early and appropriately aggressive surgical intervention has helped reduce the infant mortality in PRS and its related cleft palate syndromes from 20% to 2-3%.[2,4]

The congenital airway obstruction in PRS is worst in the newborn period and improves remarkably during early childhood. Over the first four years of age, most children exhibit "catch-up" growth of the mandible, which allows the tongue to achieve a normal position in the floor of mouth and eliminates glossoptosis. Most of this accelerated growth occurs during the first year. Therefore, when tracheostomy is required in infancy, decannulation can usually be accomplished by the age of one to three years. It is prudent to maintain the tracheostomy until after the palatoplasty procedure, as postoperative edema from closure of the very wide cleft in PRS predisposes to temporary worsening of pharyngeal airway compromise. Mandibular growth is less predictable in the other syndromes involving cleft palate and mandibular hypoplasia, such as Stickler and Nager syndromes. Mandibular distraction osteogenesis has shown promise in speeding the natural forward growth of the jaw, and has been applied as early as 14 weeks of age.[5] While this is rarely an option for management of acute airway obstruction in the infant, it may provide a means for earlier tracheostomy decannulation.

Early and appropriately aggressive surgical intervention has helped reduce the infant mortality in PRS and its related cleft palate syndromes from 20% to 2-3%.

Airway issues resurface later in childhood if velopharyngeal insufficiency and hypernasal speech necessitate pharyngeal flap surgery. After pharyngoplasty, the intentional increase in velopharyngeal airflow resistance may also translate into increased nasopharyngeal and oropharyngeal airway resistance, causing significant obstruction. Symptoms become apparent with muscular relaxation during sleep and manifest as obstructive sleep apnea.[6] This is most apt to occur when the tonsils are hypertrophic and encroach upon the lateral flap ports, and elective tonsillectomy prior to pharyngoplasty decreases the likelihood of airway obstruction in such cases.

Airway Issues in the Craniofacial Synostosis Patient

All of the craniofacial synostosis syndromes predispose children to upper airway obstruction. Crouzon, Apert and Pfeiffer syndromes are examples of these disorders in which premature fusion of the craniofacial bony sutures of the maxilla and skull base, stunts the growth of the midface, nasopharynx and oropharynx. The incidence of airway intervention in children with **craniofacial synostosis (CFS)** has been reported to be 42-70%.[1,7]

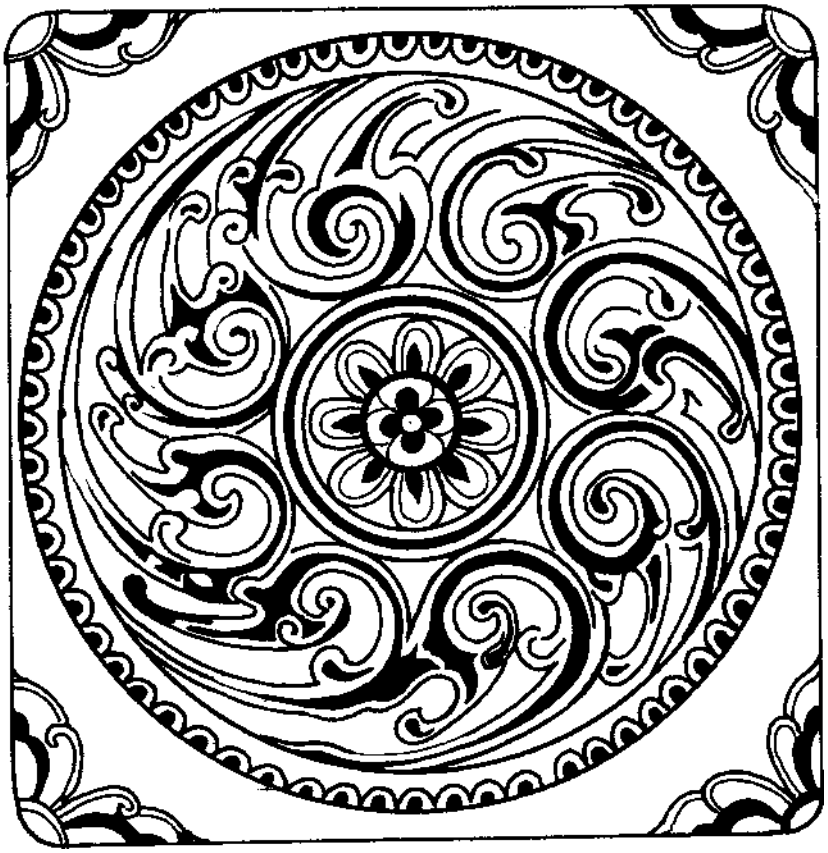
The mechanism of airway obstruction in CFS syndromes relates to arrested growth of the central craniofacial skeleton with unhindered growth of the surrounding bony and soft tissues. The typical facies of the child with Crouzon syndrome reflects the apparent retrusion of the midface as the relatively prominent frontal bones and mandible develop appropriately. Normal growth of the nasopharyngeal and oropharyngeal soft tissues gradually impinges upon the airway with even modest nasal mucosal thickening or hypertrophy of adenotonsillar tissues producing significant obstruction.

Infants with severe maxillary hypoplasia and tiny nasal cavities may present at birth with a clinical picture mimicking choanal atresia. Because they are obligate nasal breathers, affected newborns manifest stertor, retractions, cyclical cyanosis and oxygen desaturation with feeding. Catheters cannot be passed beyond the anterior nares. The diagnosis is made by high-resolution CT scan, which demonstrates stenotic nasal airways from the nostrils to the nasopharynx, with patent choanae. Initial therapy includes topical vasoconstrictors, steroid nasal drops and the avoidance of vigorous nasal suctioning to reduce mucosal edema. The narrow nasal passages do not accept a nasopharyngeal tube, but an oral airway may provide temporary relief of respiratory distress. After approximately six weeks of life, infants become better mouth breathers and the airway status may improve. If respiratory distress is refractory to these measures, tracheostomy is war-

ranted. A diagnostic bronchoscopy should be performed at the time of surgery, since children with CFS have a relatively high incidence of cartilaginous tracheal anomalies,[1,7] which may predispose to ongoing upper airway obstruction despite tracheostomy.

Surveillance for upper airway obstruction must continue through early childhood as progressive crowding of the pharyngeal soft tissues can precipitate obstructive sleep apnea. Polysomnography is a useful tool to determine the presence of sleep apnea, distinguish central from obstructive events, and to document the severity of disease. Tonsillectomy and adenoidectomy is the first surgical intervention and is most beneficial when lymphoid hyperplasia arises, typically during the period of 3 to 6 years of age. Nasal **continuous positive airway pressure (CPAP)** is tolerated by some children and may effectively eliminate obstructive sleep apnea if it persists after adenotonsillectomy. Midfacial advancement surgery improves upper airway obstruction in older children through forward distraction and expansion of the maxilla. Due to the high longterm failure rate when undertaken in early childhood, necessitating revision surgery, midfacial advancement is rarely recommended before the age of nine years. Tracheostomy is indicated for recalcitrant obstructive sleep apnea and for perioperative airway management during midfacial advancement surgery. The need for tracheostomy in children with CFS therefore has a bimodal distribution, in early infancy and at the end of the first decade of life, with an overall incidence of 33%.[1] This figure rises to 48% when mandibular hypoplasia accompanies maxillary hypoplasia, as occurs in Treacher Collins and Nager syndromes.[7] The average duration of tracheostomy is much less predictable than in PRS, since the airway in children with CFS does not uniformly improve over time.

Children with CFA are at risk for upper airway obstruction which may present with respiratory distress at birth or obstructive sleep apnea later in childhood. The trend is toward improvement in the airway with growth and development in children with PRS; the opposite is true for many patients with CFS. These children are prone to obstruction at multiple levels of the upper airway which makes airway management challenging and the incidence of tracheostomy relatively high. An understanding of the airway issues helps the primary care physician and members of the craniofacial team act together to manage airway problems in children with CFA.



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Sharon E. Gibson, MD, is Director of Pediatric Otolaryngology, Hasbro Children's Hospital; Assistant Clinical Professor, Department of Surgery, Division of Otolaryngology, Brown Medical School; and Assistant Clinical Professor, Department of Otolaryngology-Head & Neck Surgery, Tufts University.

Correspondence:
Sharon E. Gibson, MD
100 Dudley St.
Providence, RI 02905
phone: (401) 274-2300
fax: (401) 272-1302
e-mail: SGibson@lifespan.org

Anesthesia for Craniofacial Surgery

Nancy Stein, MD, and Andrew Triebwasser, MD

Of all the plastic surgical procedures, correction of craniofacial deformities in children offers perhaps the greatest challenge to anesthesiologists. Every since John Snow published the first report of giving ether to a seven year old boy for cleft lip repair in the *Lancet* in 1847,[1] anesthesiologists have been striving to perfect the safety of craniofacial operations.

In 1937, Phillip Ayre described the first use of the T-piece during cleft lip repair in infants to prevent hypercarbia, a technique that became a standard method for many years. Still, craniofacial operations carry a high risk of complications, among them venous air embolism,[2-4] difficult intubation, hypercarbia, massive exsanguination, hypothermia, cerebral edema, loss of airway, and airway obstruction.[5]

In spite of the potential problems associated with craniofacial surgery, 87% of patients report a subjective improvement in appearance;[6] 91% of parents of small children and 77% of adolescents would repeat the decision to undergo surgery.[7,8]

Surgical Considerations

Craniofacial surgery is a broad term referring to both cranial and facial surgery for the correction of cranial dysostosis or craniofacial dysmorphism. Craniosynostosis refers to premature fusion of cranial sutures, the most common form of which is scaphencephaly (saggital suture). Other recognized types include trigonocephaly (metopic), plagiocephaly (lambdoid) and brachycephaly (coronal). Early detection and surgical correction correlate with improved cosmetic outcome.

Surgical positioning varies with suture abnormalities, but can be either prone or supine. The table is rotated 90 degree. Surgical duration is in the 4-8 hour range. Mortality should be < 1-2%, with operative injury to the superior saggital sinus among the most catastrophic surgical complications. Morbidity includes meningitis, CSF rhinorrhea, intracranial hypertension, and venous thrombosis. Blindness has been reported in <1% of patients.[14]

Craniofacial deformities are associated with 58 recognized syndromes…

Pre-Operative Preparation

Craniofacial deformities are associated with 58 recognized syndromes; Apert's and Crouzon's syndrome are the most common. Associated **central nervous system** (**CNS**), pulmonary and cardiac anomalies may occur.[9] Isolated nonsyndromic craniosynostosis occurs in 6 per 10,000 births, 57% affecting the sagittal and 20% the coronal suture. Apert's syndrome occurs in approximately 1 per 100,000 births and is so severe that many of the patients require preoperative tracheostomy due to airway obstruction.

Up to 23% of craniofacial patients in one study had intracranial hypertension during sleep, even without hydrocephalus. Hydrocephalus is common, especially in Apert's and Crouzon's Syndrome, due to low cranial volumes. However, clinical signs and symptoms of elevated intracranial pressure are uncommon. Intracranial pressure has been shown to normalize two to eight weeks after surgery.

The pre-operative exam should focus on the airway, heart and lungs. 20-37% of craniofacial patients exhibit airway anomalies and, as many as 53% have intra-operative airway problems.[5,11] The old chart should be reviewed, with special reference to prior intubations, and parents advised that peri-operative airway compromise might necessitate tracheostomy. Choanal atresia, macro-glossia, micrognathia and facial asymmetry may contribute to the sudden loss of airway during induction; awake fiberoptic intubation may be required. These patients are frequently left intubated post-operatively, due to massive fluid resuscitation intra-operatively, as well as airway anomalies; arrangements for post-operative sedation and ventilation in the **pediatric intensive care unit** (**PICU**) should be anticipated.

A pre-operative history of snoring, daytime somnolence or morning headaches may be signs of nighttime sleep apnea in older children. Pre-operative sedation should be withheld unless close monitoring is feasible.

Chronic severe airway obstruction with hypoxia can lead to pulmonary artery hypertension and subsequent cor pulmonale; tracheostomy is usually performed before this condition develops.

Significant heart murmurs should be evaluated by echocardiograms pre-operatively. If an intracardiac shunt exists, air emboli may enter the coronary or cerebral circulation with catastrophic consequences.

The presence of an upper or lower respiratory infection may render an already difficult situation disasterous; thus the procedure for these children should be postponed.

The placement of central venous and arterial catheters is routine in many of these procedures, and the risks and complications of these invasive monitors should be appreciated and discussed with the parents pre-operatively.

Laboratory testing should include a hemoglobin and coagulation studies (PT, PTT, platelets) as well as a type and cross.

Anesthetic Management

Associated respiratory, cardiac, and neurological disorders will influence the anesthetic technique. Usually, an inhalation induction is

satisfactory, unless airway obstruction is deemed imminent. Awake or sedated, fiberoptic intubation in children can be difficult. A wide variety of airway equipment should be available, including **laryngeal mask airway (LMA)**, lightwand, fiberoptic laryngoscope, guidewires and, of course, the ability to perform a rapid tracheostomy.

After intubation, two large IVs and an A line are mandatory, because these procedures may involve large blood loss (as defined: greater than 20% of the blood volume). Blood loss of up to 65% of blood volume can accompany even a simple craniectomy. Longer cases may involve transfusion of greater than one blood volume ("massive transfusion"), necessitating platelets and fresh frozen plasma to prevent coagulopathy. The Hemocue® is a useful tool to monitor serial hemoglobin in these situations.

Since the procedure may last eight to ten hours, the patient's position and temperature must be carefully monitored. The wide-open cranium presents a large area for heat loss. Warm IV fluids and other peri-operative warming devices are indicated.

Routine methods of reducing cranial hypertension, such as hyperventilation, may be necessary. Although control of $PaCO_2$ and mean arterial pressure is often adequate, mannitol, furosemide, and dexamethasone should be available. Many neurosurgeons prefer that patients receive mannitol, furosemide and dexamethazone at induction.

Air embolism with circulatory collapse is a risk during craniectomy, even in the supine position. Precordial doppler monitoring is optimal,[2,3,4] but generally not utilized except in the sitting position.

Finally, severe post-operative swelling of the face often occurs after longer procedures, and post-operative ventilation and sedation may be deemed necessary to provide a safe airway until the swelling resolves.

In summary, severe complications including intraoperative death from loss of airway, massive blood loss and air embolism may occur during craniofacial surgery. Post-operative cerebral edema, brain damage and loss of airway have also been reported in the literature. At our institution, we have had no intraoperative or post operative deaths from craniofacial surgery. The anesthetic management of these specialized cases is handled by an experienced team of pediatric anesthesiologists and operating room personnel. This has led to a very low complication rate. These skilled professionals provide not only excellent perioperative care, but ensure smooth transfer to the pediatric intensive care unit for post-operative management.

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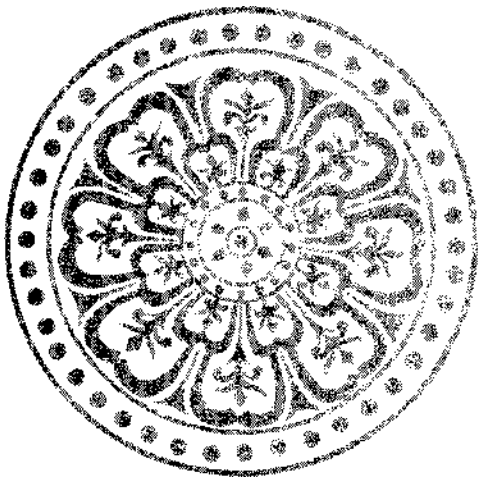
Nancy Stein, MD, is a staff anesthesiologist at Rhode Island Hospital

Andrew Triebwasser, MD, is an attending anethesiologist at Rhode Island and Clinical Assistant Professor of Anesthesiology, Brown Medical School.

Correspondence:
Nancy Stein, MD
Rhode Island Hospital
593 Eddy St.
Providence, RI 02903
phone: (401) 444-5142
fax: (401) 444-5083



CME Background Information

This CME activity is sponsored by Brown Medical School.

Target Audience
This enduring material is designed for physicians licensed in Rhode Island.

CME Objectives
After completing this CME activity, the primary care physician will be able to meet the following objectives:
At the conclusion of this course, participants should be able to:
*recognize when an infant's or child's chin, nose, and/or ears are outside normal dimensions
*detail the current understanding of craniosynostosis and positional posterior plagiocepahy; use an algorithm for the diagnosis and management of head shape abnormalities
*evaluate and manage craniofacial dysmorphology
*list the etiology of different patterns of hearing loss in children with craniofacial abnormalities
*describe the therapeutic management of speech, language, and hearing for children with cleft palates
*describe the management of airways in children with craniofacial abnormalities
*describe the anesthesia used for craniofacial surgery.

Needs Assessment
Although surgeons can correct many craniofacial anomalies, rapid detection and treatment are necessary. These papers are intended to update physicians on the diagnosis and treatment of craniofacial anomalies.

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This issue was published December 2001. This actvity is eligible for CME credit through November 2002.

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Acknowledgement
The material contained in this issue is a result of the editorial work by Patrick Sullivan, MD, guest editor. The opinions expressed herein are those of the authors and do not necessarily reflect the views of the sponsors, publisher or the planning committee.

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1. Overall quality of this CME activity	1 2 3 4 5
2. Content	1 2 3 4 5
3. Format	1 2 3 4 5
4. Faculty	1 2 3 4 5
5. Achievement of educational objectives	
* Recognize when an infant's or child's chin, nose, and/or ears are outside normal dimensions	1 2 3 4 5
* Detail the current understanding of craniosynostosis and positional posterior plagiocepahy; use an algorithm for the diagnosis and management of head shape abnormalities	1 2 3 4 5
* Evaluate and manage craniofacial dysmorphology	1 2 3 4 5
* List the etiology of different patterns of hearing loss in children with craniofacial abnormalities	1 2 3 4 5
* Describe the therapeutic management of speech, language, and hearing for children with cleft palates	1 2 3 4 5
* Describe the management of airways in children with craniofacial abnormalities	1 2 3 4 5
* Describe the anesthesia used for craniofacial surgery	1 2 3 4 5

Please comment on the impact that this CME activity might have on your practice of medicine.

_____

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Additional comments and/or suggested topics for future CME activities.

_____

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CME Craniofacial Questions

1. Prompt diagnosis and therefore early surgery in craniosynostoses can significantly improve the ultimate result by the craniofacial team. The following are all advantages to early surgery EXCEPT:
A. The calvaria bone is still malleable and therefore more easily shaped into desired form
B. Bony defects will usually ossify secondary to the rapid ossification rate
C. Releasing stenotic areas might reduce functional impairments that would otherwise possibly worsen
D. Less pshyiologic stress will be experienced by the patient and parent

2. The ideal timing of otoplasty for physical and psychological development is:
A. 2-3 years old
B. 5-6 years old
C. 9-10 years old
D. 12-14 years old

3. Rhinoplasty is performed
A. prior to the prepubertal growth spurt while tissues are more malleable
B. always through external incisions
C to achieve aesthetic goals while addressing any identified nasal obstruction component
D. always with a chin augmentation procedure

4. The basic head shape of posterior positional plagiocephaly compared to unilateral lambdoid synostosis is?
A. Oval vs circle.
B. Square vs rectangle.
C. Parallelogram vs rhomboid
D. Triangle vs circle.

5. What is the incidence craniosynostosis in the United States?
A. 1:10,000.
B. 15:10,000
C. 1:100,000
D. 15:100,000

6. Posterior positional plagiocephaly is associated with all of the following except?
A. Torticollis
B. Strabismus
C. Gene mutations
D. Head position preference

7. With regard to CS, what statement is false?
A. Mutations in growth factor receptors have been identified in both syndromic and nonsyndromic cases of CS.
B. The dysmorphic features in CS are caused by abnormal growth of the involved suture with growth remaining normal at adjacent sutures.
C. The diagnosis of CS and posterior positional plagiocephaly is primarily based on clinical examination and confirmed by radiographic studies.
D. Posterior positional plagiocephaly is due to premature fusion of the lambdoid suture.

8. In the 1850s, Virchow extensively studied the detailed growth of cranial vault sutures. He subsequently devised a law regarding the growth physiology of cranial sutures. This law states:
A. The normal growth of cranial vault sutures is parallel to the suture and in synostoses the growth is perpendicular
B. The growth of cranial vault sutures is independent of the underlying dura and the underlying brain
C. The normal growth of cranial vault sutures is perpendicular to the suture and in synostoses the growth is parallel.
D. The growth of cranial vault sutures is related to a "functional matrix" of the entire craniofacial system

9. Deformational plagiocephaly and true craniosynostoses each have defining characteristics that enable an astute physician to differentiate between the two. All of the following physical exam characteristics are seen in deformational plagiocephaly EXCEPT:
A. The superior orbital rim is depressed on the side of the flattened forehead
B. The palpebral fissure is usually oval on the side of the flattened forehead -
C. The nasal root is usually midline
D. The ear is positioned posterior and inferior on the side of the flattened forehead

10. Genioplasty is indicated to/in
A. achieve facial harmony with the nose and forehead
B. improve vertical dimension of the chin
C. adolescents with mature dentition
D. all of the above

11. By what age are the ossicles fully developed?
A. 8 weeks gestation
B. 4 months gestation
C. At birth
D. 2 years

12. What percentage of children with a cleft palate will require tympanostomy tube insertion?
A. 35%
B. 50%
C. 75%
D. 90%

13. What percentage of children with congenital deafness will have a normal temporal bone CT scan?
A. 10%
B. 15%
C. 50%
D. 85-95%

14. Children with cleft palate defect will often have hypernasality. Hypernasality is:
A. excessive nasal resonance
B. too little nasal resonance
C. articulation problems caused by pharyngeal frication
D. nasal emission on sounds such as s,z,sh, and ch

15. Conductive hearing loss is often seen in children with history of cleft palate. Children can be identified with hearing loss early using
A. pure tone audiometry
B. play audiometry
C. otoacoustic emission/auditory evoked potentials
D. tympanometry

16. A newborn with Pierre Robin sequence has intermittent stridor with retractions and poor feeding. Continuous pulse oximetry demonstrates oxygen desaturation with bottle feeds. Airway management should begin with:
A. Supplemental oxygen by nasal cannula
B. Tongue-lip adhesion procedure
C. Prone positioning and gavage feedings
D. Tracheostomy

17. The parents of a four year old child with Crouzon syndrome report he is snoring loudly, with gasping sounds and frequent awakening. Polysomnography verifies the clinical diagnosis of obstructive sleep apnea. The appropriate initial surgical intervention is:
A. Midfacial advancement
B. Tracheostomy
C. Pharyngoplasty
D. Tonsillectomy and adenoidectomy

18. A two day old with Pierre Robin sequence exhibits severe, progressive respiratory distress. Positioning, nasogastric feedings and a nasal trumpet fail to relieve stridor, marked retractions, and poor ventilation. The airway is most appropriately managed by:
a. Tracheostomy
b. Long term endotracheal intubation
c. Hyomandibulopexy
d. Mandibular distraction osteogenesis

19. All of the following are feared complications during craniofacial surgery except
A. venous air embolism
B. saggital sinus tear
C. loss of airway
D. bradycardia due to oculocardiac reflex

20. Which monitor is not of value in rapid diagnosis of air embolus?
A. end-tidal carbon dioxide monitoring
B. EEG
C. precordial doppler
D. pulse oximetry

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Atrial Fibrillation and Anticoagulation

Robert Maxim, MD

There are 1.8 to 2.2 million people with atrial fibrillation in the United States. Atrial fibrillation accounts for one third of all admissions to hospitals for dysrhythmia. The median age for someone with atrial fibrillation is 75. One of 20 patients over age 69 will have atrial fibrillation. This increases to 1 of 11 for those >80 years old.[5] The overall rate of stroke among those with atrial fibrillation is 4.5% per year and is as high as 8.1% in high- risk individuals. Anticoagulation with warfarin reduces this risk by more than two thirds. The risk for major bleeding is 1.3% with warfarin and 1% with aspirin. The higher an individual's risk for stroke the more benefit warfarin conveys. Despite this evidence warfarin continues to be underutilized especially in elderly patients who are most at risk for stroke.[3,4]

Who Should Be Treated With Warfarin?

Table 1 describes the recommended treatment for patients with atrial fibrillation according to risk factors. Patients with any risk factors, or those over the age of 75 with atrial fibrillation should be treated with warfarin. Either aspirin or warfarin may be considered in those 65-75 years old with no additional risk factors (CHF, HTN, prior stroke or TIA). Aspirin or no treatment can be considered in those under 65 years old without any additional risk factors.[3]

Hypertension and age provide the greatest increase in risk for stroke in those patients with atrial fibrillation. Hypertension alone is responsible for an absolute risk of 3.7%/year, whereas the absolute risk for stroke is 3.5%/year for those over 75 years of age. The presence of multiple risk factors increase absolute risk of stroke by between 5.0 and 7.0%/year.[1]

How Well Do The Results Of Randomized Clinical Trials Correlate With Actual Clinical Practice?

A recent comparison of trials involving patients with atrial fibrillation in **actual clinical practice** conditions (**ACP**) and **randomized controlled trials** (**RCT**) showed an increase in minor bleeding in ACP. More importantly the rates for major bleeding in ACP (1.1%) were similar to RCT (1.3%) and for ischemic stroke (1.8% vs 1.4%).[6]

What Are The Treatment Goals for Anticoagulation With Warfarin?

For most patients on warfarin a target INR of between 2.0 to 3.0 results in a reduction in the relative risk for stoke by as much as 68%. Anticoagulation in this range raises the absolute risk for serious hemorrhage by 0.3%/year. However, close monitoring of anticoagulation is necessary because of a narrow therapeutic window. Anticoagulation clinics have been demonstrated to be an effective method of maintaining INRs in an effective range.

Is Aspirin Effective In Reducing Stroke In Patients With Atrial Fibrillation?

For patients at low risk for stroke, aspirin alone is recommended; and for those with a contraindictaion to warfarin, aspirin (325mg/d) is a modestly effective alternative. Aspirin alone reduces the relative risk for stroke by 21% compared to 68% with warfarin. The relative risk for major bleeding with aspirin is increased by 0.5% per year.

Table 1
Recommended Treatment for Patients with Nonvalvular Atrial Fibrtlation

Age	Risk Factor	Absolute Risk	Recommended Treatment
>75	none	3.5%/y	warfarin
>75	one or more	8.1%/y	warfarin
65-75	none	4.3%/y	warfarin or aspirin
65-75	one or more	5.7%/y	warfarin
<65	none	1.4%/y	aspirin or none
<65	one or more	4.9%/y	warfarin

Risk Factors

Hypertension	Prosthetic heart valve
Transient ischemic attack (TIA)	Rheumatic mitral valve disease
History of stroke or systemic embolus	Poor left ventricular function

Should Warfarin And Aspirin Therapy Be Combined?

Studies comparing fixed dose warfarin and aspirin to adjusted dose warfarin alone have shown a higher rate of stroke in those on combination therapy. An increase in yearly stroke rate of 7.9% was noted for those on combination therapy versus 1.9% for adjusted dose warfarin.[3] Combined therapy is not advised.

Who Is At Increased Risk For Bleeding?

There is no medical evidence to support the common belief that falls in older patients

Table 2

Outpatient Bleeding Risk Index[2]

Risk Factor	Relative Risk of Major Bleeding	Index Points
Age 65 or greater	3.2	1
History of: Recent MI, Cr> 1.5, Hct <30%, or Diabetes Mellitus	3.0	1
History of stroke	2.5	1
History of GI bleed	2.9	1

Estimation of Risk

Level of Risk	Sum of Index Points	Major Bleeding (%/yr)
Low Risk	0	3
Intermediate Risk	1-2	12
High Risk	3-4	48

are a significant cause of intracranial bleeding. Patients most likely to experience intracranial hemorrhage are those with established ischemic cerebrovascular disease and hypertension. The risk for major bleeding can be estimated by using the Outpatient Bleeding Risk Index (Table 2). A sum of index points based on independent risk factors can categorize a patient as low, intermediate, or high risk for major bleeding. This index can help predict risk for major bleeding when combined with other pertinent clinical information.[2] Evidence suggests that there is an increased risk for intracranial hemorrhage in patients over the age of 75 as the INR exceeds 2.5. In this population a lower INR (target range of 1.6 to 2.5) could be considered. It is important to note that maintaining control of the INR in target range is the best approach to reducing the risk for bleeding or subtherapeutic complications.

What Can I Do To Reduce My Patient's Risk Of Bleeding?

Just as subtherapeutic anticoagulation (INR< 1.5) decreases the effectiveness of warfarin, supratherapeutic anticoagulation increases the risk of bleeding. Active patient involvement in the treatment process correlates with adequate anticoagulation. Patients aware of their INR results are more likely to be adequately anticoagulated.[7] Age is not a risk factor for poor anticoagulant control.[8]

Evidence suggests control of both systolic and diastolic blood pressure may reduce the risk of bleeding. A review of 5 randomized controlled trials showed those patients who experienced intracranial hemorrhage had a higher average systolic and diastolic blood pressure than those who did not.[9] Additionally, the **Systolic Hypertension in the Elderly Program (SHEP)** trial demonstrated that stroke was reduced by 36% by adequate control of blood pressure.[10]

Conclusion

Physicians should consider anticoagulation with warfarin in all their patients with atrial fibrillation evaluating both new and previously diagnosed patients not on warfarin for their risk of stroke.

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Raymond Maxim, MD, is Clinical Coordinator at RIQP; and staff physician at Roger Williams Medical Center.

Correspondence:
Raymond Maxim, MD
Phone (401) 528-3265
Fax: (401) 528-3210
e-mail: ripro.rmaxim@sdps.org

6SOW-RI-AFIB-01-03

The analyses upon which this publication is based were performed under Contract Number 500-99-RI02, entitled "Utilization and Quality Control Peer Review Organization for the State of Rhode Island," sponsored by the Health Care Financing Administration, Department of Health and Human Services. The content of this publication does not necessarily reflect the views or policies of the Department of Health and Human Services, nor does mention of trade names, commercial products, or organizations imply endorsement by the U.S. Government.

The author assumes full responsibility for the accuracy and completeness of the ideas presented. This article is a direct result of the Health Care Quality Improvement Program initiated by the Health Care Financing Administration, which has encouraged identification of quality improvement projects derived from analysis of patterns of care, and therefore required no special funding on the part of this Contractor. Ideas and contributions to the author concerning experience in engaging with issues presented are welcomed.

Health by Numbers

Rhode Island Department of Health
Patricia A. Nolan, MD, MPH, Director of Health

Edited by Jay S. Buechner, PhD

Utilization of Inpatient Rehabilitation Services

Jay S. Buechner, PhD, and Edward F. Donnelly, RN, MPH

For many injuries and diseases, treatment in the acute care setting is only the first stage in the patient's return to good health and normal functioning, and is followed by inpatient and/or outpatient rehabilitation treatment. This subsequent period may last longer and cost more than the acute care phase, and the insurance coverage of the health care costs incurred may not be as complete as it is for acute care. Rehabilitation care also presents different challenges in evaluating the quality of care and the outcomes of care than are presented by acute care. This analysis briefly presents descriptive data on inpatient rehabilitation treatment in Rhode Island, including characteristics of the treated patients and patterns of their care; there is no equivalent source of data for outpatient rehabilitation care.

Methods

Since October 1, 1989, all acute-care general hospitals in Rhode Island have submitted patient-level data for every hospital inpatient stay, as required by licensure regulations. As of October 1, 1998, the discharge data reporting requirements were extended to the state's two psychiatric specialty hospitals and one inpatient rehabilitation facility. Under these requirements, up to eleven diagnoses made during the hospital admission are included as codes from the **International Classification of Diseases, 9th Revision, Clinical Modification (ICD-9-CM)**.[1] For this analysis, cases of rehabilitation treatment were defined as either (1) any patient discharged from the rehabilitation facility or (2) any patient discharged from one of the acute care general hospitals with a principal diagnosis of "Care involving use of rehabilitation procedures" (ICD-9-CM code V57). The analysis included all such discharges during the twelve-month period from October 1, 1999, through September 30, 2000, corresponding to hospital fiscal year 2000.

Information on the medical condition underlying the patient's need for rehabilitation was drawn from the reported diagnosis codes. For patients of the inpatient rehabilitation facility, the information was drawn from the principal diagnosis; for patients of the acute-care general hospitals, the information was drawn from the first-listed additional diagnosis, as the principal "diagnosis" for those patients was the need for rehabilitative care.

For the purpose of computing average lengths of hospital stays, patients not staying overnight were counted as having a stay of one day, per industry practice.

Results

During the twelve-month period investigated, there were 2,260 discharges of patients whose hospital stay was for rehabilitation. Of these, 776 (34.3%) were from the state's only licensed inpatient rehabilitation facility, the **Rehabilitation Hospital of Rhode Island (RHRI)**. The remaining 1,484 were treated in acute care facilities, nearly all of them in four hospitals, Kent County Memorial Hospital, the Memorial Hospital of Rhode Island, Newport Hospital, and St. Joseph Hospital. Rehabilitation patients represented 1.3% of all (non-newborn) patients in the state's acute care hospitals, but between 2.5% and 6.1% of patients in the four hospitals where they were concentrated.

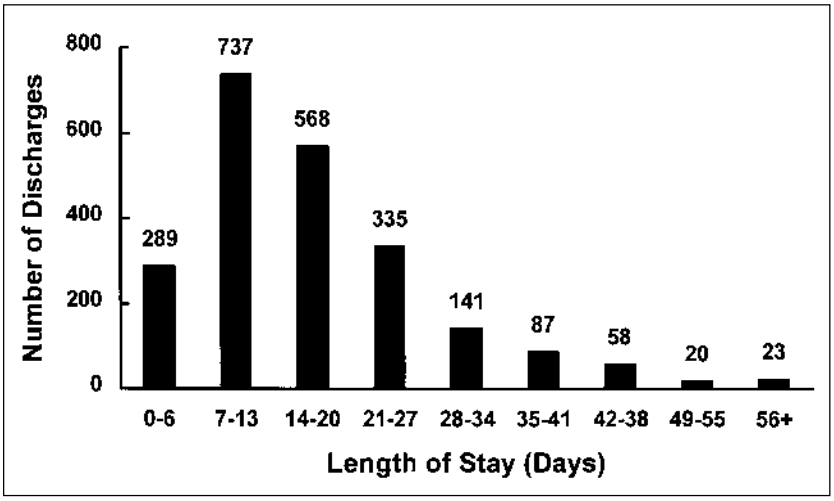

Figure 1. Length of Stay among Inpatient Rehabilitation Patients, Rhode Island, October 1, 1999 - September 30, 2000.

Rehabilitation patients generally experienced longer hospital stays than most acute care patients. For all facilities combined, the **average length of stay (ALOS)** for rehabilitation patients was 17.1 days. By facility, ALOS for rehabilitation patients ranged from 12.5 days to 19.4 days. The statewide median and mode (most

Table 1. Medical Conditions among Inpatient Rehabilitation Patients, Rhode Island, October 1, 1999 – September 30, 2000.

Medical Condition	Number of Discharges	Percent of Discharges
Diseases of the circulatory system	627	27.8
Injury and poisoning	447	19.8
Diseases of the musculoskeletal system and connective tissue	266	11.8
Diseases of the respiratory system	190	8.4
Diseases of the nervous system and sense organs	187	8.3
Symptoms, signs, and ill-defined conditions	129	5.7
Diseases of the genitourinary system	81	3.6
Endocrine, nutritional and metabolic diseases, and immunity disorders	79	3.5
Neoplasms	65	2.9
Diseases of the blood and blood-forming organs	26	1.2
Diseases of the digestive system	22	1.0
Diseases of the skin and subcutaneous tissue	22	1.0
Mental disorders	11	0.5
Infectious and parasitic diseases	10	0.4
Congenital anomalies	7	0.3
Complications of pregnancy, childbirth, and the puerperium	1	0.0
Certain conditions originating in the perinatal period	0	0
Supplemental classifications	89	3.9

frequently occurring) values for length of stay were both 14 days. The distribution of length of stay for these patients shows that a sizable proportion (14.6%) of patients had stays extending to four weeks and beyond; the longest reported stay was 117 days. (Figure 1)

The need for inpatient rehabilitation treatment was driven by a wide range of medical conditions. The largest single group of conditions among these patients was diseases of the circulatory system (27.8%), followed by injuries and poisonings (19.8%), musculoskeletal conditions (11.8%), respiratory conditions (8.4%), and diseases of the nervous system (8.3%). (Table 1)

The age distribution of rehabilitation patients was heavily skewed toward higher ages, with the large majority of patients being ages 55 and older (84.5%). More specifically, 12.2% were ages 55-64, 25.8% were 65-74, and 46.5%, or nearly half, were 75 years old or older. The majority of patients were females, who comprised 57.8% of all patients and 62.3% of those ages 65 and older.

A considerable majority (63.5%) of inpatient rehabilitation patients were covered by Medicare, as would be expected from the age distribution of these patients. (Figure 2) The balance were covered primarily by various private health plans (29.2%), with smaller proportions covered by Medicaid (4.8%) and Workers' Compensation (1.0%). Only 1.5% of these patients were uninsured for their care.

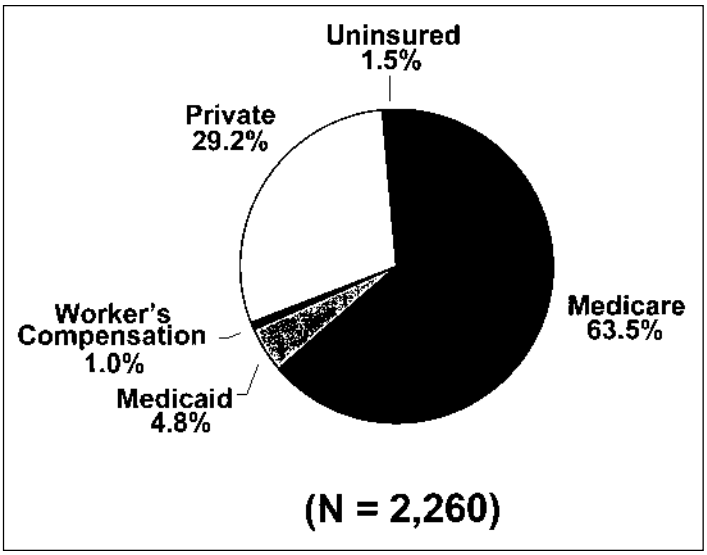

Figure 2. Expected Source of Payment among Inpatient Rehabilitation Patients, Rhode Island, October 1, 1999 - September 30, 2000.

Discussion

With the inclusion of the state's single inpatient rehabilitation facility, the statewide hospital discharge data system now covers all providers of inpatient rehabilitation care in the state. Based on these data, a majority of such care is provided by the acute care hospitals, although the Rehabilitation Hospital of Rhode Island is the largest single provider of care. The average stay for rehabilitation care is over two weeks, but many patients stay for one month and longer. The typical patient is elderly, is covered by Medicare, and is most likely to be recovering from a chronic disease of the circulatory, musculoskeletal, respiratory, or nervous system or from an injury.

Rehabilitation treatment is part of a continuum of care; it usually follows a period of acute care and may in turn be followed by long-term care, such as in a nursing home or from a home care provider. The capacity to link or otherwise combine information on patients across these settings is useful to estimate the costs of medical care for specific preventable conditions or to evaluate the efficacy of the health care system in ultimately returning patients to good health and functioning. The establishment of an information source for rehabilitation care provided by inpatient facilities in our state is an important step in building this capacity.

Jay S. Buechner, PhD, is Chief, Office of Health Statistics, and Assistant Professor of Community Health, Brown Medical School.

Edward F. Donnelly, RN, MPH, is Senior Public Health Epidemiologist, Office of Health Statistics and Clinical Teaching Associate, Department of Community Health, Brown Medical School.

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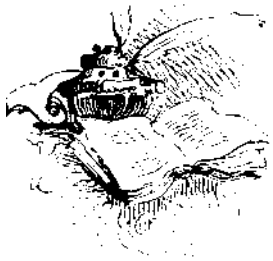
– A Physician's Lexicon –



Aks Me No Questions

Some medical terms regularly convey a chilling, despairing sense of foreboding. One of these disquieting words is metastasis, derived from the Greek, *meta-* , meaning next to, beyond, changed or transformed; and *-stasis* , meaning place, location or the sense of immobility. Physicians have taken this word, originally defined neutrally as the removal of something to another site, and have narrowed its definition to the process of depositing cancer cells to sites physically removed from the primary neoplasm.

The *-stasis* root appears in many medical terms, often denoting stoppage as well as immobility, as in hemo*stasis*: it may also appear in the form of *-stat* in words such as thermostat, prostate [to stand before the bladder], bacteriostatic, orthostatic, or even ecstatic [ec- , in Greek, meaning away from; hence a rapturous feeling akin to being transported from the immobile].

The *meta-* [sometimes *met-*] root is also found in numerous medical terms. Metacarpal/metatarsal define bones which are next to the wrist [Greek, *karpos*] or the foot [Greek, *tarsos*]. *Meta*chromasia describes the process whereby a cell assumes a color other than the color the dye staining it. Metabolism denotes the totality of energy producing/consuming chemical changes within living tissue. Methemoglobin is a transformed form of oxyhemoglobin; and yet other words such such as metaphase, metanephros, metaplasia, metanephrine and metaphysis repeat the root.

There is also metaphor [the application of a word to an object which it does not literally define; a figure of speech]; metamorphosis [*morpho-* , meaning form, thus a transformation of a structure into something else]; metaphysics [currently thought of as studies beyond the realm of physics]; and metathesis, that wondrous historic act of transposing or displacing letters or syllables within words. Thus, when people utter such allegedly unlettered pronunciations as *childern*, *prespiration*, *modren* or *govrenment* they are not being perversely ignorant; they are merely - and probably unknowingly - enunciating the Old English pronunciations of these words.

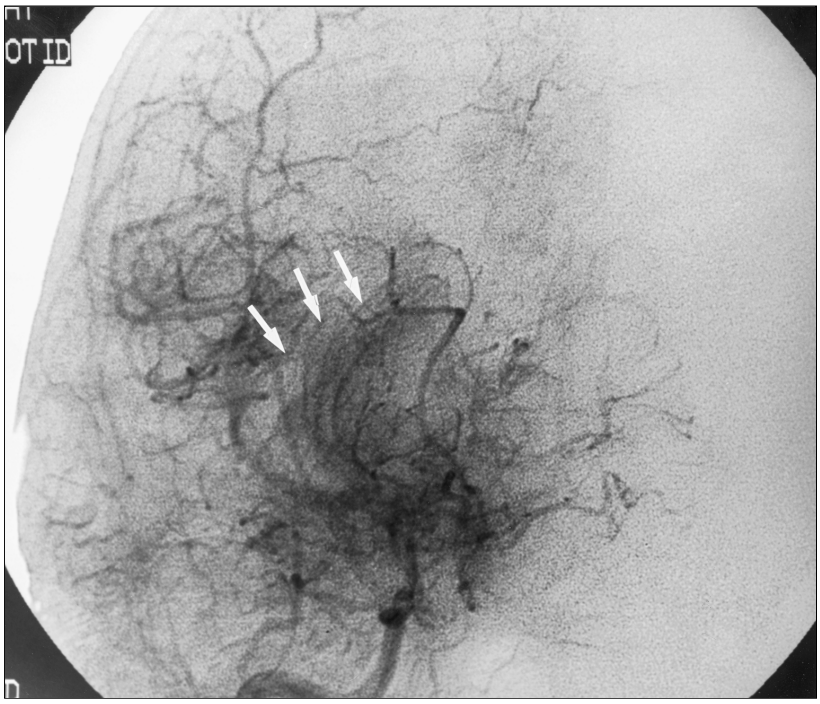
When innocent children therefore say, "May I aks a question?" adults should not respond with righteous indignation since the Old English verb meaning to ask, was indeed *aksien*. And some day Scriptural scholars may return, without wincing, to "Aks, and it shall be given to you; seek and ye shall find; knock and it shall be opened unto you."

Given a choice, metathesis is a much more congenial word than metastasis.

– Stanely M. Aronson, MD, MPH

IMAGES IN MEDICINE

Edited by John Pezzullo, MD



Moyamoya

A 42 year-old female with a prior history of stroke at age 28 presented with acute onset of left hand and leg weakness. CT scan of the brain (not shown) revealed an old left frontal infarct, and an acute hemorrhage in the right basal ganglia. The history of recurrent stroke in a young female suggests an underlying vasculopathy or vascular malformation in addition to cardioembolic disease. Cerebral angiography (right carotid injection shown) performed shortly after admission demonstrated severe cerebrovascular-occlusive disease involving the anterior circulation with the development of a rich collateral network and dilatation of the lenticulostriate vessels. The result is the pathognomic "puff of smoke" on cerebral angiography (arrows) characteristic of Moyamoya.

Moyamoya is a rare chronic cerebrovascular disorder of unknown etiology characterized by progressive stenosis of the intracranial carotid arteries as well as the Circle of Willis. As a result, a rich collateral circulation forms from extracranial-intracranial anastomoses, and dilatation of the lenticulostriate vessels. The distended lenticulostriates are responsible for the "puff of smoke" appearance on cerebral angiography. There are two peaks in incidence – children under ten years who present with ischemic infarct, and adults in the third or fourth decade who present with hemorrhagic stroke. Treatment includes surgical direct or indirect revascularization such as superficial temporal artery- middle cerebral artery anastomoses, and encephalomyosynangiosis.

JAMES Y. CHEN, RICHARD L. GOLD, MD, PHILIP B. VAIDYAN, MD

James Y. Chen is a 4th year medical student at Brown Medical School.
Richard L. Gold, M.D. is Radiologist-in-Chief at the Miriam Hospital.
Philip B. Vaidyan, M.D. is a Clinical Instructor of Internal Medicine, Brown Medical School.

CORRESPONDENCE:
Philip B. Vaidyan, M.D.
Medicine Associates
phone: (401) 331-8555
e-mail: PVaidyan@lifespan.org

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Images in Medicine: We encourage submission to the Images in Medicine section from all medical disciplines. Image(s) should capture the essence of how a diagnosis is established, and include a brief discussion of the disease process. The manuscript should be less than 250 words and include one reference. The manuscript and one or two cropped 5 by 7 inch prints should be submitted with the author's name, degree, institution and e-mail address to: John Pezzullo, MD, Department of Radiology, Rhode Island Hospital, 593 Eddy St., Providence, RI 02903. An electronic version of the text should be sent to the editor at jpezzullo@lifespan.org.

Cutaneous Anthrax

Laura E. Regan, MD, Leslie Robinson-Bostom, MD, Martin A. Weinstock, MD, PhD

B. Anthracis is a large gram positive nonmotile spore-forming bacterial rod. The spores are infectious and may persist for years in the environment. Outside of the context of bioterrorism they are most commonly found in hides and grazing areas of infected animals. From 1950 through 1979, there were 443 cases of anthrax in the United States; from 1980 through 1999, there were 6.[1,2] Infection can occur after inhalation, ingestion or cutaneous contact with anthrax. Previously, cases in the United States were limited almost exclusively to workers in the animal product industry, particularly workers handling hides. In Rhode Island the last case occurred in 1969.[3,4]

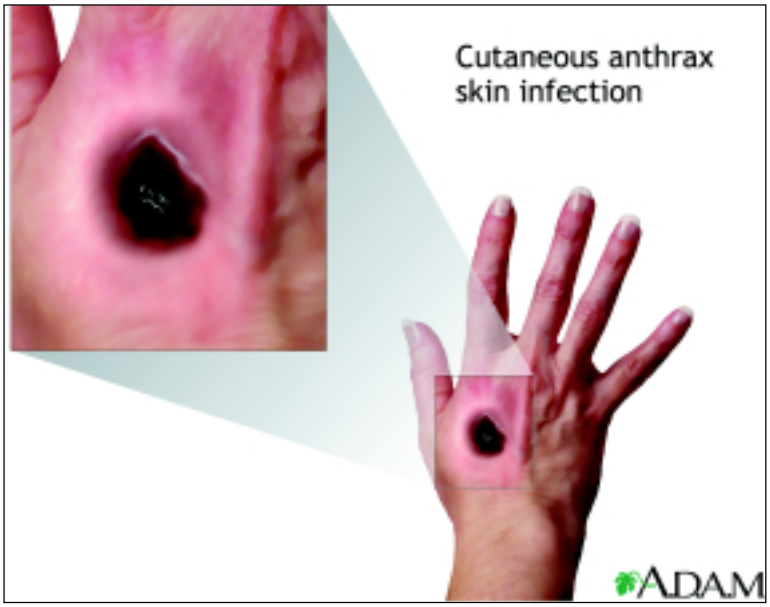
B. Anthracis cannot penetrate unbroken skin. After a one to twelve day incubation period, the patient may experience nonspecific symptoms of fever and malaise. In more than 95% of cases a lesion (sometimes referred to as a "malignant pustule") forms at the site of contact, most commonly on the extremities. Initial clinical appearance is a small asymptomatic or pruritic macule or papule. This slowly enlarges to form a bullous lesion with surrounding erythema and edema. These may became hemorrhagic and appear necrotic. Satellite lesions may be noted. A characteristic black eschar (the word anthrax derives from the Greek word for coal) forms at the site. Lesions are typically painless, and may be accompanied by regional adenopathy.[5,6]

If you suspect anthrax , notify the Rhode Island Department of Health immediately, prior to collection of cultures and specimens. During regular office hours call (401) 222-2577, after hours (401) 272-5952. If the patient has samples of the possible agent, call the local police (911) as well.

Laboratory evaluation includes gram stain and culture of cutaneous lesions. When vesicles are present, cultures of the fluid of a previously intact vesicle are recommended. During the eschar stage, eschar should be gently lifted and swabs rotated underneath the eschar for cultures.

Two 4-mm punch biopsies are also recommended, one for culture and one for hematoxylin and eosin permanent sections. Biopsy is recommended of the edge of the lesion if the vesicular stage is present, and of the erythematous area next to the eschar if the eschar stage is present. If both manifestations are present, specimens from both areas should be obtained. The specimen for permanent section should be placed in room temperature formalin. The culture sample should be sent in a sterile container with sterile saline directly to the microbiology laboratory. The local laboratory should be alerted beforehand to the suspected anthrax. In addition to gram stain and culture, fungal and atypical mycobacterial cultures should be performed. The laboratory should be asked to reserve a small specimen of fresh tissue at -70 degrees centigrade for possible future PCR testing. A detailed clinical history should accompany each specimen.

Two 5 ml red topped tubes of blood should be obtained. One tube will be used for ELISA testing, and should be labeled "Rule out Anthrax" and stored at -70 degrees centigrade. The second tube will be used for PCR testing by the Rhode Island Health De-



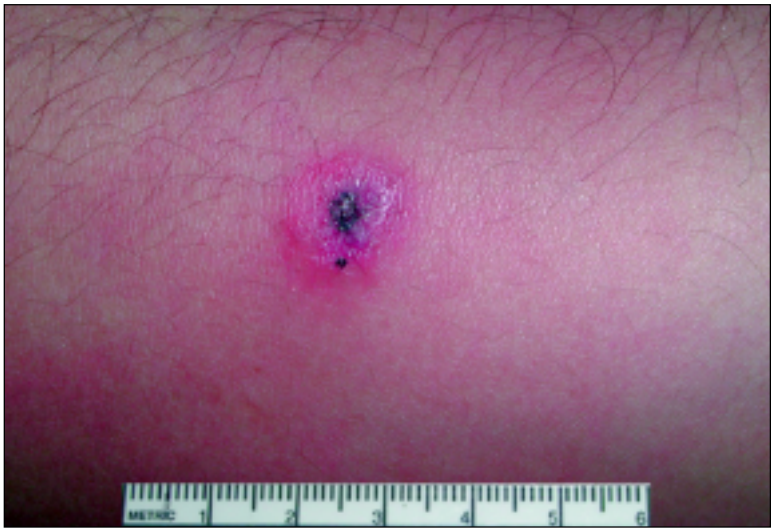


Table 1. Treatment for cutaneous anthrax patients without systemic symptoms and not located on the head or neck and not with extensive edema

Category	Initial Oral Therapy	Duration
Adults	Ciprofloxacin 500 mg BID **OR** Doxycycline 100 mg BID	60 days[a]
Children	Ciprofloxacin 10-15 mg/kg every 12 hours (not to exceed 1g/day)[b] **OR** Doxycycline:[c] > 8 years and > 45 kg: 100 mg every 12 hours All other children: 2.2 mg/kg every 12 hours	60 days[a]
Pregnant Women[d]	Ciprofloxacin 500 mg BID **OR** Doxycycline 100 mg BID	60 days[a]
Immuno-compromised persons	Same as for non-immunocompromised persons and children	

a: previous guidelines have suggested 7-14 days, but 60 days is recommended in the setting of this attack, given the likelihood of simultaneous exposure to aerosolized *B. anthracis*
b: Ciprofloxacin or doxycycline should be considered first-line therapy. Amoxicillin 500 mg po TID for adults or 80 mg/kg/day divided every 8 hours for children is an option for completion of therapy after clinical improvement.
c: The American Academy of Pediatrics recommends treatment of young children with tetracyclnes for serious infections (e.g., Rocky Mountain spotted fever).
d: Although tetracyclines or ciprofloxacin are not recommended during pregnancy, their use may be indicated by life-threatening illness. Adverse effects on developing teeth and bones are dose-related; therefore, doxycycline might be used for a short time (7-14 days), before 6 months of gestation.

Table 2. Treatment of cutaneous anthrax patient with systemic symptoms or extensive edema or involving the head or neck (same as for inhalational anthrax exposure)

Category	Intravenous Therapy[e]	Duration
Adults	Ciprofloxacin 400 mg every 12 hours **OR** Doxycycline 100 mg every 12 hours **AND** one or two additional antimicrobials[e]	IV treatment initially.[f] Switch to antimicrobial therapy when clinically appropriate. Continue for 60 days (IV and po combined).[a]
Children	Ciprofloxacin 10-15 mg/kg every 12 hours (not to exceed 1g/day)[g] **OR** Doxycycline[c]: >8 years and >45 kg: 100 mg every 12 hours All others: 2.2 mg/kg every 12 hours **AND** one or two additional antimicrobials[e]	IV treatment initially.[f] Switch to oral antimicrobial therapy when clinically appropriate. Continue for 60 days (IV and po combined).[a]
Pregnant Women[d]	Same as for non-pregnant adults. The high death rate from the infection outweighs the risk posed by the antimicrobial agent.	IV treatment initially.[f] Switch to oral antimicrobial therapy when appropriate. Continue for 60 days (IV and po combined).[a]
Immuno-compromised Persons	Same as for non-immunocompromised.	Same as for non-immuno-compromised.

e: Other agents with *in vitro* activity include rimfampin, vancomycin, penicillin, ampicillin, chloramphenicol, imipenem, clindamycin, clarithromycin. Because of concerns of constitutive and inducible beta-lactamases in *B. anthracis*, penicillin and ampicillin should not be used alone. Consultation with an infectious disease specialist is advised.
f: Intial therapy may be altered based on clinical course of the patient; one or two antimicrobial agents may be adequate as the patient improves.
g: If intravenous ciprofloxacin is not available, oral may be acceptable because it is rapidly and well absorbed from the gastrointestinal tract with no substantial loss by first-pass metabolism.

Tables 1 and 2. Reproduced with modifications from the American Academy of Dermatology Bioterrorism Task Force web site: http://www.aad.org/BioInfo/Biomessage2.html. verified 11/14/01.

partment and needs refrigeration. If patients are febrile or hospitalized, blood cultures are recommended.[7]

Patients with suspected anthrax should be started on the appropriate antibiotics immediately and should continue for 60 days unless anthrax is ruled out. Tables 1 and 2 list recommended treatments. [7,8] Because these recommendations may change, clinicians should consult these sources:

*http://www.healthri.com (RI Department of Health)

*http://www.CDC.gov (Centers for Disease Control and Prevention)

*http://www.aad.org (American Academy of Dermatology Bioterrorism Task Force)

Additional information may be found in references 9 and 10 and at *http://www.tray.dermatology.uiowa.edu

Acknowledgements

The authors thank Dr. Louis Fragola and Suleka Neelgaru for their assistance.

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Laura E. Regan, MD, is a resident in the Department of Dermatology, Brown Medical School.

Leslie Robinson-Bostom, MD, is an Assistant Professor and Director, Division of Dermatopathology, Department of Dermatology, Brown Medical School.

Martin A. Weinstock, MD, PhD, is Chief of Dermatology, VA Medical Center, Providence; Director, Photomedicine Division, Pigmented Lesion Unit, and Dermatoepidemiology Unit, Rhode Island Hospital, and Professor, Department of Dermatology, Brown Medical School.

Vital Statistics

Rhode Island Department of Health
Patricia A. Nolan, MD, MPH, Director of Health

Edited by Roberta A. Chevoya

Rhode Island Monthly Vital Statistics Report

Provisional Occurrence Data from the Division of Vital Records

Underlying Cause of Death	Reporting Period			
	December 2000	12 Months Ending with December 2000		
	Number (a)	Number (a)	Rates (b)	YPLL (c)
Diseases of the Heart	278	3,109	314.5	4,089.0 **
Malignant Neoplasms	202	2,431	245.9	7,020.0
Cerebrovascular Diseases	54	508	51.4	782.5
Injuries (Accident/Suicide/Homicide)	50	362	36.6	6,757.5
COPD	48	507	51.3	397.5

Vital Events	Reporting Period		
	June 2001	12 Months Ending with June 2001	
	Number	Number	Rates
Live Births	933	13,264	13.4*
Deaths	782	10,209	10.3*
Infant Deaths	(10)	(100)	7.5#
Neonatal deaths	(10)	(85)	6.4#
Marriages	980	8,550	8.6*
Divorces	227	3,228	3.3*
Induced Terminations	505	5,519	416.1#
Spontaneous Fetal Deaths	60	995	75.0#
Under 20 weeks gestation	(49)	(920)	69.4#
20+ weeks gestation	(11)	(75)	5.7#

(a) Cause of death statistics were derived from the underlying cause of death reported by physicians on death certificates.

(b) Rates per 100,000 estimated population of 988,480

(c) Years of Potential Life Lost (YPLL)

Note: Totals represent vital events which occurred in Rhode Island for the reporting periods listed above. Monthly provisional totals should be analyzed with caution because the numbers may be small and subject to seasonal variation.

* Rates per 1,000 estimated population # Rates per 1,000 live births
** Excludes one death of unknowqn age.

THE RHODE ISLAND MEDICAL JOURNAL

The Official Organ of the Rhode Island Medical Society
Issued Monthly under the direction of the Publications Committee

VOLUME 1
NUMBER 1

PROVIDENCE, R.I., JANUARY, 1917

PER YEAR $2.00
SINGLE COPY, 25 CENTS

Ninety Years Ago

[December, 1911]

Fredric J. Farnell, MD, from Butler Hospital, contributed "The Wasserman Reaction - Its Significance in Nervous and Mental Diseases." He cautioned, "It should be clear....all obscure nervous and mental affections which show a positive serum reaction are not necessarily of a syphilitic genesis." Dr. Farnell cited Dr. Plaut, a pioneer in clinicopathological psychiatry, who had "studied the serum and spinal fluid in all types of nervous and mental disease." In 187 cases, he found serum positive for all but 3 (1 cerebrospinal syphilitic and 2 tabetics), the spinal fluid positive in all but 30 (6 general paralytics, 14 cerebrospinal syphlitics and 10 tabetics).

In "Chronic Catarrahal Colitis," Charles F. Peckham, MD, noted that many people have a weak colon, "like a weak throat or a weak heart." He described the reputation of the colon as marginal: "From the literature we have been led to believe that the colon is an organ well-nigh devoid of function. That in the domestic economy of modern life it had, as its sole usage, the storage of feces. ... it has been placed in the same category as the appendix - among the relics of a more specialized ancestry." For colonic disturbances, Dr. Peckham advised patients to eliminate alcohol and tobacco, eat more fruits and vegetables, less fat.

John R. Elliott, MD, in "Empyema," described the 12 cases he had seen in four months as house-surgeon at Rhode Island Hospital. He recommended urotropin for patients with pneumonia (50 grams daily for adults - lower doses were "utterly valueless.") All 12 cases were treated by "simply opening into the pleural cavity between two ribs in the axillary or post-axillary line and draining with...tubes.....resection of a portion of the rib is the only proper treatment."

Fifty Years Ago

[December, 1951]

Ivan Basylewycz, MD, on the Medical Staff of Rhode Island State Hospital, formerly Professor of Medicine, Kiev, Ukraine, contributed "Pernicious Anemia in Old Age." For this disease, "A therapeutic test of liver treatment should be carried out in all doubtful cases."

Thad A. Krolicki, MD, Assistant Surgeon, Proctology, Memorial Hospital, contributed "Rectal Bleeding." He urged physicians to consider any adult patient with a history of bleeding from the rectum to have a malignancy "until proved otherwise."

Alexander A. Jaworski, MD, and John E. Farley, MD (residents in pediatrics at the Charles V. Chapin Hospital), contributed "Congenital Heart Block - Review and Report of a Case of Congenital Complete Heart Block with Rheumatic Fever, and Physiological Studies." This 12 year-old boy, born at home, was admitted with Vincent's angina when he was 14 months old. His pulse was 70 per minute. At age 22 months, his pulse was 88; two weeks later, his pulse was 60; at age 4, his pulse was 50. At age 12, he was feeling fine, with no symptoms, but for three days developed pain in his ankles and feet. Subsequently he was diagnosed with bradycardia. The authors conclude, "The prognosis for a normal life span in the patient is guarded."

Twenty Five Years Ago

[December, 1976]

Stanley S. Freedman, MD, in "The Early Practice of Allergy in Rhode Island," began with Dr. Maruice Adelman's establishment of a clinic for asthmatic children at Rhode Island Hospital in 1924. "Pediatricians took turns willingly donating their time and effort to the clinic." That early clinic led to a formal Pediatric Allergy Clinic, still functioning in 1976. Staff attended monthly Boston meetings of the SWI (Sneeze, Wheeze and Itch) club. In 1938 Rhode Island Hospital established an Adult Allergy Clinic (Dr. Clinton Westcott, Chair, Department of Medicine, RIH, "would not allow the tiny, though vital, Department of Pediatrics to outdo the august Department of Medicine.") Francis Chafee was the first director of the adult clinic.

Thomas J. Paolino, MD, in "A Review and Comparison of the Psychoanalytic and Sociological Complications of the Alcoholic Marriage," declared that "Research which blends the sociological and psychodynamic factors is long overdue."

A.A. Savastano, MD, and Joseph B. Fitzgerald, MD, in "School Screening for Spinal Deformities in Rhode Island," advocated a "program…to prevent unnecessary pain and permanent deformity." In 1959 Dr. Savastano and Dr. John Thayer had established a Scoliosis Clinic at RIH; by 1976 the clinic was meeting twice a week. In 1974-5 school personnel and the medical team, after screening 7,699 Rhode Island students, identified 160 with visual evidence of spinal deformity; they were advised to see their physician. Of the 93 who saw physicians, 60 were treated.

In "Message from the Dean," Stanley M. Aronson, MD, commented on the Health Manpower Bill (Health Professional Education Assistance Act of 1976). The bill would establish a National Health Service corps, set federal criteria for determining allowable increases in college tuition, require residencies to provide "a reasonable number" of shared-schedule rotations, institute a program of federally insured loans for students, end preferential visa status for noncitizen Foreign Medical Graduates (since physicians were no longer considered a shortage), and require schools to set aside for American foreign medical students a certain number of advanced-standing class positions, assigned by the Secretary of Health Education and Welfare. The Message questioned the need for such federal intrusion: Brown regularly enrolled Americans who had studied at foreign universities, providing them with a year of supervised clinical experience and a Modified Fifth Pathway Program.