

PEDIATRIC NEWS

San Antonio Military Pediatric Center



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Housestaff Puzzler

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L is a 19-year-old G1P1 female who presented to the Darnell Army Community Hospital (DACH) at 2300 complaining, "my head hurts so bad I feel like I am dying." She reported waking that morning with a pounding bilateral frontal headache, and that the pain had progressively worsened throughout the day; she stated that the pain was currently 10/10, and that the only thing that made it better was lying down in the dark. She denied visual changes, nausea, vomiting, neck stiffness, fever, previous severe headaches, weakness or sensory changes in her extremities, edema, trauma, drug abuse, depression/anxiety, ataxia, vertigo, rashes, cough, urinary symptoms, vaginal discharge or bleeding, or other symptoms.

Her past medical history was significant for giving birth to a healthy infant term female

Continued on page 10

In this edition

| | |
|--|---|
| Housestaff Puzzler | 1 |
| Pyloric Stenosis in the 21st Century .. | 1 |
| Neonatology for the General Pediatrician | 1 |
| Sickle Anemia: An Update | 6 |
| Professionalism in Patient Care: Tradition and Change | 8 |

Pyloric Stenosis in the 21st Century

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Pediatricians and surgeons will encounter infants with pyloric stenosis. The incidence is about 1 per 250 live births. There is a male predominance. A familial predisposition has also been described especially in first-born male infants.¹

Pyloric stenosis was originally described at autopsy more than 100 years ago by the Danish pediatrician Harald Hirschsprung. Most of these children died with this disease secondary to severe dehydration, electrolyte abnormalities, and cardiovascular collapse. Ramstedt described the classic operative therapy of pyloric stenosis in 1911. The fundamentals of Ramstedt's approach are still used today to successfully treat pyloric stenosis.²

After Ramstedt and Fredet described the Pyloromyotomy, surgical correction of this disorder became the mainstay. In all major series in the literature including a review of over 1500 cases at Los Angeles Children's Hospital, mortality is nonexistent and morbidity is extremely low.³

Currently, the major innovation in the treatment of pyloric stenosis has been in the minimally invasive technique. Tan in 1993 described the first laparoscopic technique, which has been modified only slightly. The advantages of this technique are superior cosmetic result and a lower wound infection rate.⁴

This article will cover the salient points of the presentation, examination, diagnosis, and operation. A clear understanding of pathophysiology and treatment of pyloric stenosis is paramount to maintaining the standard of care.

Continued on page 2

Neonatology for the General Pediatrician

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What follows is a discussion of the recommended follow-up care and evaluation schedule for the Neonatal Intensive Care Unit graduate. The focus will be on what the general Pediatrician will be confronted with in the out patient setting.

Very Low Birth Weight (VLBW) infants account for approximately 1.4% of the 3.4 million annual US births. Over the last few decades there has been an increase in the survival

Continued on page 4

Presentation

Pyloric stenosis classically presents at 4-6 weeks of life with nonbilious emesis. The emesis is often described as “projectile”, “poltergeist”, or “hitting the wall”. Some infants have significant dehydration with altered mental status or lethargy. The child may have a sunken fontanelle, dry mucous membranes and decreased urination. Other infants will present more insidiously with formula intolerance having gone through several feeding changes with minimal benefit. Some infants will even have failure to thrive as an initial diagnosis. In reality, infants present within a spectrum from severe dehydration to formula intolerance. The sine qua non of pyloric stenosis, however, is nonbilious emesis.

Examination

If you can palpate the pyloric tumor, then you have made the diagnosis with expert certainty. Examination is a dying art in modern medicine but the superior clinician will follow these steps. First, the clinician should always go to the operating room with the attending surgeon on these cases to palpate the “olive” (pyloric tumor) while the child is under general anesthesia. This experience will allow one to “see” with their fingers. The old adage is true that if you have never “seen” (palpated) it then you are unlikely to know what you are looking for (or palpating). At teaching institutions, residents, housestaff, etc are welcome in the operating room to experience the controlled exam.

Preoperatively, in the clinic or ER, palpation of the olive requires two things: 1) patience and 2) more patience. There are several tricks in

the examination. The child should be lying supine on the exam table. If the stomach is distended, then a nasogastric or orogastric tube is helpful. The infant will also be more cooperative if you let him/her nipple 1/2 ounce of dextrose water while performing the exam. The examiner should be on the child’s right side. Sometimes, 15-20 minutes is required to get a good exam. Patience. Another important aspect of the exam is knowledge and identification of known structures on the abdominal exam such as the spine, kidneys, psoas and liver edge.

Diagnosis

To make the diagnosis requires only one of three possible tests. First, a reliable exam with palpation of an “olive” is definitive. If the examination is inconclusive then an ultrasound should be obtained.

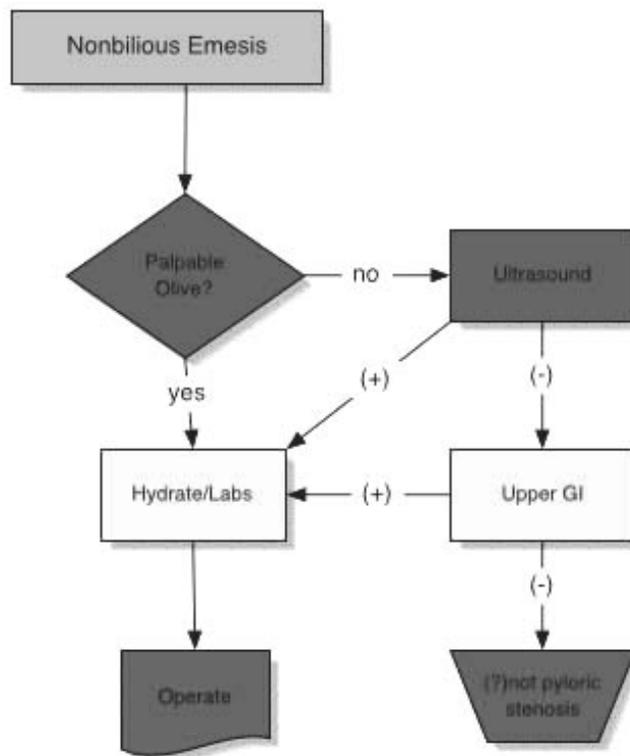
and the channel length is ≥ 16 mm.¹ If ultrasound is indeterminate or unavailable then an upper GI can be obtained. The upper GI should show a string sign through the pylorus, “shouldering”, and/or peristaltic waves. Contrast must pass through the pylorus before definitive diagnosis can be made.

Preoperative Care

Infants with pyloric stenosis all have some degree of dehydration. The child should be made NPO. A complete blood count and chemistry panel should also be obtained. IV fluids (D₅1/2NS) are started. KCl should be added after the child urinates. Several questions arise from the statements above:

1. Why do we use D₅1/2NS and not D₅1/4NS or something else?

Generally, pyloric stenosis causes a global chloride deficit from



A simple diagnostic approach to pyloric stenosis

protracted vomiting. 1/2NS replaces the lost chloride quicker than 1/4NS.

2. How long do you hydrate the child before surgery and why?

Children with pyloric stenosis are intravascularly dry – sometimes very dry. They have a significant metabolic alkalosis. Intravascularly dry infants who undergo general anesthesia will have a transient decrease in vascular tone, which can result in cardiovascular collapse (and death). Typically, a chemistry panel is checked every 8 hours until the serum bicarbonate is less than 30. When the serum bicarbonate is less than 30, then the infant is hydrated enough to have general anesthesia.

3. Why add KCl and why wait until the infant urinates?

The classic metabolic abnormality in pyloric stenosis is a hypokalemic hypochloremic metabolic alkalosis. Occasionally, these infants are so profoundly dehydrated that they are in oliguric renal failure. Potassium replacement could cause hyperkalemia and the resultant cardiac arrhythmias.

4. Another common question relating to the urinalysis is why do some babies have a paradoxical aciduria?

Physiologically, humans with a metabolic alkalosis should have alkaline urine. The opposite is also true. If a person has an acidosis then the urine should have an acid pH. However, infants with pyloric stenosis have a metabolic alkalosis and sometimes a paradoxical aciduria. Why? The Na⁺-Proton exchanger in the proximal tubule initially conserves Na⁺ and dumps K⁺. This explains the infant becoming hypokalemic. The urine is alkaline. However, once the degree of serum hypokalemia reaches a

certain point then the Na⁺-Proton exchanger switches to excreting a H⁺ (instead of K⁺) for each Na⁺ ion that is conserved. The urine then becomes paradoxically acid despite a systemic alkalosis.

Operative Therapy

The fundamentals of the operation have remained essentially unchanged for nearly 100 years. The pyloric channel must be “opened”. This is achieved by splitting the hypertrophied pyloric muscle down to the mucosa – a Pyloromyotomy. The pyloric channel is able to open up and allow passage of stomach chyme. Recent modifications to the operation have been on the approach through the abdominal wall to the pylorus.

The standard skin incision for pyloric stenosis is made halfway between the umbilicus and the xiphoid and centered just to the right of midline. This incision is directly over the pylorus and allows for optimal exposure. The drawback of this incision is the relatively large scar. Because of this cosmesis issue, two other approaches have developed.

The omega incision is an incision made around the top curve of the umbilicus and extended on both sides if necessary to make an omega sign. The incision is brought down to the fascia and then a tunneling technique is used to expose the fascia superiorly. The fascia is opened and the pylorus brought out through the incision. This approach has two disadvantages. One there is an increased incidence of incisional hernias. Second, sometimes the pyloric tumor is so large that exposure is difficult. The major advantage of this method is the cosmesis of the incision, which is hidden, in the umbilical fold.

The laparoscopic approach offers superior cosmesis and exposure. There have also been reports of decreased times to the

resumption of full feeds and lower incidence of vomiting. An incision is made at the umbilicus for the camera. Also two other tiny incisions are made in the abdominal wall just below the costal margins on the right and left side. Statistical significance will require larger numbers to validate these findings. However, the growing trend in pediatric surgery is toward the laparoscopic approach.

Postoperative care

Infants with pyloric stenosis recover quickly from the operation. IV fluids are continued until full feeds are tolerated. Feeds are usually started 6 hours postoperatively. The most common feeding schedule is as follows:

15 cc of pedialyte followed 3 hours later by:

30 cc of pedialyte followed 3 hours later by:

30 cc of formula followed 3 hours later by:

45 cc of formula followed 3 hours later by:

60 cc of formula followed 3 hours later by:

formula ad lib

This regimen works well unless the infant vomits. If vomiting occurs then feeds are continued in 3 hours at the same volume. Only after this volume is tolerated, the feeds are increased. If a child vomits twice on consecutive feeds then the child should be made NPO and feeds restarted in 6-8 hours.

Pain control is usually achieved with rectal Tylenol for 24 hours. The incisions are infiltrated at the time of operation with local anesthetic (usually marcaine). Narcotic analgesics are not indicated. These infants are all less than 60 weeks gestation and need apnea/bradycardia monitoring for 24 hours post-operation.

Complications

Complications occur in 1-2% of cases. The most common complication is wound infection. Wound infections are usually managed by opening the wound and doing dressing changes. Second most common complication is perforation of the pylorus intraoperatively. This requires a stitch to repair the injury. These infants will not start feeds for 24 hours. Another complication is incomplete pyloromyotomy. This complication requires a another operation. Usually this is discovered by continued vomiting. Upper GI is used to demonstrate the persistent nature of the pyloric stenosis. The pylorus is split in a spot 120-180 degrees from the original incision. Other complications are much less common such as bowel injury, bleeding, or sepsis.

Conclusion

Pyloric stenosis is a modern surgical success story. Diagnosis should be made on physical exam, but can be confirmed by ultrasound. Preoperative hydration of these infants is crucial. Operative therapy is moving toward the umbilical omega incision or the laparoscopic approach. These techniques offer primarily improved cosmesis.

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Neonatology

Continued from page 1

rates of VLBW infants with survival being reported in as low a gestational age as 22-23 weeks. Current NICHD data report survival of approximately 30% at 23 completed weeks gestation with an increase to 80% survival by 25 weeks. With increased survival of these small infants, there has been an increase in the morbidities associated with prematurity. Chronic lung disease, developmental delays, retinopathy of prematurity, strabismus, amblyopia, refractive errors, hydrocephalus, short gut syndrome, cerebral palsy, hearing loss, and other significant disabilities may be seen in Neonatal Intensive Care Unit survivors. Up to 10% of overall survivors will have a severe disability or handicap at school age. Of the remainder, many will have serious difficulties secondary to mild delays. In addition, there are often psychosocial factors which contribute to the infants overall outcome. The primary care Pediatrician must be able to recognized and deal with the myriad of issues seen in these patients.

The first step in the continuing care of the premature infant after discharge is the initial meeting of the family with the primary care Pediatrician. Whenever possible, the Pediatrician should meet with the family and see the patient while he or she is still in the Neonatal Intensive Care Unit. This allows the Pediatrician to establish a bond with the family and to get a baseline exam on

the infant. If this is not possible, phone contact can be made and an appointment for initial evaluation should be made for as soon as possible after discharge. The timing of this first appointment will be dependent upon the physical status of the infant and the psychosocial needs of the family.

After discharge, the Pediatrician must establish a follow-up schedule with the family. Frequently, these visits need to be more frequent than would seem to be medically indicated. Often, families may feel insecure in their abilities to care for their premature infants. They have just left a very structured and well-monitored situation for the relative “comfort” of home. They may have underlying fears and need time to adjust to the home situation. The level of attention can be decreased after the parents have had time to deal with their anxieties. They need to know that their worries are not trivialized.

The Pediatrician must also be aware of the special needs of each infant. There are several common concerns but each infant will have their own unique needs. They may be on home monitoring equipment, oxygen or ventilators. They may have feeding tubes and feeding pumps. Nurses may be coming to the home. The Pediatrician must be familiar with all of the equipment used and the local administrative procedures for obtaining and maintaining this equipment.

The first common concerns are the growth and developmental status of the infant. These parameters should be assessed using an adjusted age scale. To obtain an adjusted age, you take the chronologic age and subtract the weeks or months of prematurity. Growth parameters can be plotted on adjusted age graphs or on standard growth charts. However, if using standard charts, the plotting should be at the adjusted age with a notation explaining this. Premature

infants tend to leave the hospital in a relatively growth retarded state. They also tend to gain weight at a slower rate than their age adjusted peers. Expected weight gain should be at least 20 to 25 g/day for up to 3 months adjusted age and 10 to 20 g/day from 3-6 months adjusted age. Significant illness, i.e. chronic lung disease, can significantly increase the caloric needs of these infants as well. Even with out significant illness, however, premature infants will most likely need additional nutritional support to establish this expected growth. Studies have shown that infant's discharged on special formulas will have improved growth parameters. In addition, breast milk alone may not meet the premature infant's nutritional needs and supplementation may be necessary. There are commercial products available that can be added to breast milk or fed as a supplement after breast feeding. Special premature formulas are also available that are designed to meet the increased nutritional needs of those infants that are formula fed. Supplemental nutrition may be recommended up to 1 year but may only be needed for the first 3-6 months.

Developmental issues should be assessed at regular intervals with enough time between each evaluation to allow for expected developmental progress. These evaluations should begin with an assessment of risk factors by the Pediatrician. Knowing the medical history, interviewing the parents, using standardized screening tools such as the Ages & Stages Questionnaire and making repeated clinical observations over time are the recommended steps for screening. Infants at risk and those with identified problems should be referred for a formal developmental evaluation. Many recommend starting at approximately 3 to 6 months adjusted age and them following every 3 to 6 months to assess for continued developmental

progress. Blindness, hearing impairment, and cerebral palsy can be diagnosed during the first 18 months to 2 years. Intellectual deficits may not be clearly diagnosed until the later preschool years or early school-age years. Functional learning impairments and other learning disabilities may not be apparent until after the child's entrance into elementary school.

Vision and hearing should also be screened in all premature infants. Retinopathy of prematurity (ROP) affects approximately 35% of infants with birth weights of ≤ 1250 g. This increases to 60% for those weighing ≤ 750 g at birth. ROP may lead to refractive errors, amblyopia, strabismus, or blindness. Infants should receive their initial exam at 4-6 weeks of chronologic age or 31 to 33 weeks postconceptional age. These exams should be done by an ophthalmologist familiar with the retinal changes associated with this condition. Follow up exam interval will be determined by the ophthalmologist. The infant will be followed until the retinas are fully vascularized and/or any retinopathy has resolved or become stable. Infants with and without a history of retinopathy should routinely be scheduled for follow-up exams at 6 months and 1 year. After 1 year, exams should be scheduled at regular intervals to monitor for strabismus and amblyopia, which may occur at later ages. Age 3 years and just before school age is one recommended schedule.

The incidence of hearing loss in the preterm infant population is reported to be between 2% to 6%. Universal screening of all newborns is now recommended and should be done according to developmental age. Otoacoustic Emissions should be done in the first 6 months adjusted age. It is a test of peripheral function (cochlea and outer hair cells). Brainstem Auditory Evoked Response should be done within the first 3 to 6 months adjusted age. This records stimulus-based electrical

activity from the subcortical auditory structures. Older infants and children can be screened using Behavioral Audiometry. Infants should receive their initial evaluation prior to hospital discharge if possible or as soon as possible thereafter. Those that fail screening tests should be referred for confirmatory testing within 6 to 8 weeks. If indicated, hearing aid evaluations should be done prior to 6 months chronologic age.

Other outpatient screening studies may be indicated based on the infant's ongoing medical conditions. Drug levels should be done as with older children on certain medications. Electrocardiograms should be done on infants with bronchopulmonary dysplasia, especially those on long-term oxygen therapy. These infants are prone to right heart strain. These may be done up to monthly depending on the infants lung disease. An echocardiogram may also be indicated. Electrolytes should be done on infants receiving chronic diuretic therapy. They should be followed every 2 weeks until the dosage is stable. After this time, repeat screen should be done when clinically indicated. Head ultrasound, magnetic resonance imaging or computed tomography may be indicated in those infants with unresolved central nervous system issues. Hemoglobin and Hematocrit should be followed for several weeks after discharge. Adequacy of iron intake should also be confirmed. Nephrocalcinosis screening should be done in infants on chronic diuretics (dipstick urine) and finally osteopenia screening should be done in any extremely-low-birth-weight infant.

The final common point I would like to discuss is the administration of immunizations. Former premature infants should receive full doses of the routine vaccines at the appropriate chronologic age. In addition, influenza vaccine should be given to

any high-risk infant. RSV prophylaxis should also be given to at risk infants.

This has been a general review of the care of former premature infants. For a more details, the reader is referred to the references listed below.

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Sickle Anemia An Update

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Introduction

Since our first documented case of sickle cell anemia in 1910 by Herrick, we continue to see significant reductions in morbidity and mortality associated with our understanding of the pathogenesis of sickle cell anemia. Neonatal screening, penicillin prophylaxis, health maintenance and parental education have lead to longer survival, lessening in chronic end-organ damage and an improved quality of life for children and adults with sickle cell disease. With improved survival, we must address the unique areas of organ dysfunction associated with vaso-occlusive crises of the different organ systems

Pathogenesis

Sickle cell disease is a congeni-

tal inheritance of various abnormal hemoglobins leading to homozygous hemoglobin S (Hgb SS), or heterozygous forms of hemoglobin S with hemoglobin C (Sickle C disease) or hemoglobin S with beta thalassemia (Sickle thal disease). Normal red blood cells are biconcave and consist of three forms of hemoglobin (Hgb A, A2 and F). A single amino acid substitution of valine for glutamic acid on the 6th position of the beta globin chain causes the hemoglobin molecule to polymerize, form long tactoids and ultimately distorts the shape and function of the red cell (sickle shape) with deoxygenation. Recurrent sickling and unsickling causes red cell membrane damage and along with changes in blood viscosity leads to the clinical manifestation we see in sickle cell disease. Increased red cell fragility, adherence of red cells to the endothelium and phagocytes result in hemolytic anemia; increase infections and ischemic damage to various organ systems. While deoxyxgenation serves as the primary stimulus for polymerization, several other factors influence sickling to include high Hgb S concentrations, fever and cold temperatures, lactic acidosis from poor tissue perfusion and respiratory acidosis associated with sedation and dehydration. While hypoxia merits oxygen supplementation, routine oxygen is not necessary and can be harmful.

Acute Chest Syndrome

Acute Chest Syndrome (ACS) is a heterogeneous pulmonic process associated with sickle cell syndromes. It consists of a new pulmonary infiltrate in combination with a fever, chest pain and in older children, dyspnea, cough and tachypnea. Donald Rucknagel in his article "Progress and prospects for acute chest syndrome of sickle cell anemia" describes intrapulmonary vascular thrombosis as the primary cause of ACS. This can be compli-

cated by hypoventilation associated with sedation and excessive hydration. Wheezing occurs in all age groups. 70% of patients can be hypoxic (pulse oximetry < 90% or PO₂<80 mm Hg). Patients can have decreasing hemoglobin and an increase in nucleated red blood cells. Treatment is supportive with oxygen supplementation, antibiotics and possible transfusion of red cells. Physicians caring for large populations of sickle hemoglobinopathies have noted an increase in the prevalence of asthma. Leong et al in their evaluation of airway hyperactivity in children with sickle cell anemia found 83 % of patients with SCD and asthma had bronchial hyperreactivity. 64% of patients with SCD without asthma had bronchial hyperreactivity. The overall prevalence was 73%, much higher than in the general population. As with asthma, glucocorticoids may have some efficacy in management of acute chest syndrome. Steroids impair cytokine release during ischemia, alter arachidonic acid metabolism and inhibit phospholipase A₂. Clinical trials with dexamethasone at 0.3-mg/kg q 12 hours appear to shorten hospitalizations and decrease oxygen, opioid and blood transfusion requirements. Incentive spirometry, red blood cell transfusion and hydroxyurea are strategies with potential benefit in ACS.

The Role of Doppler Flow Studies in Sickle Cell Anemia

Strokes are acute neurological events caused by stenosis or occlusion of large cerebral arteries like the internal carotids and the middle cerebral vessels. Symptomatic ischemic events occur in 7-11 % of children with Hgb SS disease. MRI /MRA are diagnostic and chronic transfusion therapy appears to reduce the recurrence of strokes. Silent infarcts occur in 17 % of children between 6-12 years of age.

Transcranial Doppler ultrasonography offers a non-invasive means of detecting patients at increased risk for stroke based on increased blood flow velocities. High blood flow velocity correlate with stenosis and subsequent risk for strokes. In Adams et al study, patients were randomized to receive standard treatment or blood transfusions to maintain Hgb S concentrations less than 30%. This study suggests transfusions reduce the risk of first strokes in children with sickle cell disease and abnormal TCD. Standardization of the TCD procedure was and remains a critical determining factor in use of the data. The use of TCD to direct the need for chronic transfusion therapy and the associated hemosiderosis has an impact. We will find ourselves revisiting the need for stem cell transplant secondary to iron overload. Data from other studies suggest that children in the untreated group had stroke risk of 10% per year. Eleven strokes in the untreated group compared to one in the transfused group has led to termination of the study and recommendations for transfusion by the National Heart, Lung and Blood Institute. The use of hydroxyurea in the treatment of stroke prevention has not been fully evaluated. A study by Ware et al proposes benefit with only 19 % recurrence incidence but larger controlled studies are needed.

Hydroxyurea

Hydroxyurea is an antineoplastic agent that inhibits DNA synthesis and repair. Its use in sickle cell disease includes use for acute chest syndrome, as an alternative for chronic transfusion therapy and for use in vaso-occlusive crises. The proposed clinical benefit of hydroxyurea in sickle cell disease relate to its augmentation of fetal hemoglobin, its reduction in white cell counts, its impact on red cell deformity and altered expression of

adhesive receptors on red cells. Common protocols initiate treatment at 15 mgs/kg and increase at a rate of 5mgs/kg to a maximum of 30mgs/kg. The efficacy of hydroxyurea in adults appears to extend to children of all ages with notable improvement in hospital admissions, total hospital days and reduced transfusion requirements. Toxicity is minimal with myelosuppression being the most common effect. Its potential role in various aspects of sickle cell disease are many and most importantly will impact on the reducing chronic organ damage.

Stem Cell Transplantation

Bone marrow transplantation is the only curative measure for sickle cell disease. HLA -identical donations allow for successful replacement of donor red cells or mixed chimerism; correcting complication of sickle cell disease. Survival post – transplantation is as high as 90% with 85 % event-free survival. Graft rejection occurs in 11% of patients and death in only 8% of patients. Indications for transplantation include primary CVA, recurrent acute chest syndrome, recurrent pain crises, osteonecrosis and ocular retinopathy. Potential donors include related sibling with or without sickle trait. Allogeneic stem cell transplant can be achieved with myelablative or non-myeloablative therapy. While stem cell transplants offer curative intent and excellent long-term survival, limitations in potential related donor matches and optimal timing for transplantation (before major end organ damage of the lungs or heart, or hepatitis) remain a problem.

Summary

Advances in management techniques for sickle cell anemia continue to improve survival.

Associated complications of potential areas of end organ ischemia have been met with new and intriguing approaches to treatment. Growing alternative to management such as anti- platelet agents, thrombolytics and even baby aspirin may hold promises for the future.

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National Child Abuse Prevention Month

Professionalism in Patient Care: Tradition and Change

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Introduction

There is no ages-long tradition of the healing trades as professions, in the same sense as priestly work or, perhaps, lawyering and soldierly leadership. Even the practitioners of Hippocratic medicine, despite their pledge to abide by a set of ethical behaviors, were not professionals in the eyes of their contemporaries and did not claim that distinction. They were simply members of one of many traditions of healing, none of which rose to the societal level of profession. The ethnographic study of pre-literate and prehistoric healers seems to show that they, too, were not (or are not, in some isolated societies) elevated to the status of professionals.(1)

What is the definition of a profession? Why does it hold such sway over our self-identification as physicians? Why does it seem so important that we continue to hold a professional role in our community? The answers to the first question are generally offered in social and political terms.(2) But the answers to the latter questions are also both very personal and intimately shared, through the common experiences that bind us together as clinicians.

I will present definitions of professions and of professional behavior. I will also discuss the recently published Charter on Medical Professionalism.(3) Finally, I will suggest some possible future directions for professionalism, particularly in pediatrics.

Professions and Professional Behavior

Plumbing is not a profession. Neither is astronomy. Astrology, on the other hand, sometimes has met the definition of a profession (millennia before the Reagan administration). What is the difference?

A profession is an occupation or trade whose work is critical to the culture, which has been given the right by society to control its own work in the application of its specific knowledge and skills. It is defined more by the way it employs its knowledge and skills than by their actual nature or complexity.(2)

In return for the right to independent control over their own training, decision making, accreditation, and discipline, the members of a profession agree to place the needs of every client they serve before their own personal needs and goals. The status of profession is granted by the society itself, usually by its most powerful and influential members.(2) This is an important point, because these persons tend to be most interested in legitimizing and stabilizing the social structure, and their interests do not always parallel those of the rest of the population.

Plumbing is usually fairly simple work, but it also can be technically demanding and even risky. Nevertheless, society does not require that the plumber put the needs of a home owner with a backed-up toilet before his own. Few of us could ever master the science and skills of modern astronomy. But, like a tradesman, the astronomer works under no social expectations that he or she must rush to the aid of a father who can't identify the North Star for his child.

On the other hand, in societies where astrologers and prophets were important in religion or in political decisions, they would have enjoyed a sort of professional status in return for honest interpretation and advice,

whether it was what the client wished to hear or not. The truth of the client's situation came before the prophet's personal wishes—and, unfortunately for some seers, their personal safety.

In the United States, “scientific” physicians, as opposed to herbalists, homeopaths, and homespun healers, did not achieve professional status until the second half of the 19th century.(4) The path to professional recognition included the national organization of physicians in the American Medical Association, their insistence on stiffened criteria of medical school accreditation and physician licensure, and the triumph of scientific research over other approaches to health and illness. Each of these steps was political and social, as well as medical.

The pressure within medicine for these changes was based, in part, on a heightened sense of responsibility for the health of their fellow citizens and the determination to put the needs of the stricken before their own. But, as Starr has pointed out, there is also little question that the political and social struggle to emerge as “the” medical profession also yielded very favorable outcomes in terms of income and prestige for physicians individually and as a group.(4)

One critical aspect of professional status is almost never discussed. If a society can grant professional status to a trade or occupation, it can also remove that status if the trade fails to meet society's expectations.(2) That is, the social contract between a profession and the society in which it practices is never one-sided. Along with the privileges granted to practitioners, and the special status given to the needs of their clients, there are also attitudes and behaviors expected from both parties. If the situation of either party to the contract changes enough, it may become necessary to renegotiate the

relationship. Society can even terminate the profession's status.

Professionalism's Importance

Why is it important to us, as physicians, to think of ourselves, and to be thought of by others, as professionals? There are a number of reasons, both pragmatic and philosophical. In addition, there are as many personal reasons as there are practitioners. Pragmatically, the complexity of our knowledge and the nature of our work are best supported and protected by our professional status. Our knowledge continues to be difficult to master and to maintain. Even the increased sophistication of patients and the broad availability of medical information on the Internet are insufficient to permit patients to make well-informed decisions in all instances. Also, as Freidson has described, physicians are legally and socially the professionals through whom all health care flows, including that provided by the members of other professions, such as nursing and social work.(2) The physician is traditionally the Captain of the Ship in patient care. Even our colleagues, advanced practice nurses and physician assistants, practice within the framework of a professional relationship led by a physician.

Philosophically, the basis of our ethical conduct as physicians is the professional model. We are to put the needs of our patient before our own. The four principles of medical ethics, advocated by Beauchamp and Childress, emphasize this: respect for persons; beneficence; nonmaleficence; and justice.(5) Because of our special position in society, we must use our knowledge and skills in ways that uphold these principles.

Almost all of us also have deep personal convictions about our role as healers. It may not be possible to truly practice medicine without such convictions. In a study of family

practice residents at UCSF, Saba found that, even in the 1980s and 1990s, 85 percent of all residents identified a sense of calling or mission as a principle reason for their being physicians.(6) Since our system of health care encourages a sense of physician arrogance(7), we may need such grounding in personal conviction to practice ethically.

The Charter on Medical Professionalism

In March 2002, the members of the Medical Professionalism Project published the Charter on Medical Professionalism, which appeared simultaneously in *The Lancet* and the *Annals of Internal Medicine*.(3) The publication capped several years of work by representatives of the American Board of Internal Medicine Foundation, the American College of Physicians-American Society of Internal Medicine Foundation, and the European Federation of Internal Medicine.

“*Professionalism is the basis of medicine's contract with society.*” is the first sentence of the Charter, and could serve as its entire message. Three principles follow: the principle of primacy of patient welfare; the principle of patient autonomy; and the principle of social justice. Beauchamp and Childress's four principles fit completely into these.

The authors then present a set of professional responsibilities, defined by ten commitments that each physician should personally accept. These flesh out the principles and include, for example, a commitment to professional competence, a commitment to honesty with patients, and a commitment to professional responsibilities.

It is important to note that the authors of this Charter were all Europeans and North Americans. The social contract that the Charter describes may not be completely applicable in areas of the world with

different social and healing traditions.

Future of Professionalism.

It is not at all clear that our current definition of professionalism will remain valid. As society changes, so do its needs and its citizens' expectations of each other. I offer three suggestions regarding professionalism's future in medicine.

First, it is likely that the role of the physician will change and, with it, the concept of professional behavior. This seems inevitable, since neither our society nor the professional organization of medicine remain the same.

Second, we physicians will have to pay increasing attention to what our patients expect of us, in their own definition of what constitutes professional behavior. In part, this comes from the increasing availability of understandable medical information. It also reflects growing societal concerns about the just distribution of health resources. In addition, although this is anecdotal so far, many patients seem to be ready to take on third party payers on their own behalf. It is possible that they will view us as less and less effective guides through the maze of justification and evidence that employers and insurers demand before paying for care.

Finally, professional behavior will remain a key element in the care we provide our patients. Because of our own attitudes toward our calling or mission to care, because of our increasing voice in support of the members of society whose health needs are far from being met, because of the intensely intimate and humane nature of what we do, we will always find strength in a sense of our own professionalism.

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Housestaff Puzzler
Continued from page 1

one-week prior via C-section for failure to progress. She had had frequent prenatal visits, with no problems prior to delivery. After delivery, she was treated with IV antibiotics for postpartum endometritis, for which she was still on tetracycline for outpatient treatment. No personal or family history of hypertension, pre-eclampsia, eclampsia, migraines/headaches, brain tumors, psychiatric disorders, glaucoma, or subarachnoid hemorrhages. They had a gas furnace and working CO monitor in the home.

Physical Exam

Vitals: HR 80, RR 18, BP 190/115, SaO2 100% RA.

Gen: Obese Hispanic female lying flat on her back with the light off, moaning and holding her head; her husband was anxiously sitting next to her.

Head: NCAT, no TTP

Neck: No LAD, supple, FROM

Ears: TMs pearly

Eyes: PERRLA, no papilledema, disc margins sharp

Nose: Clear nares, no drainage, no sinus TTP

OP: No lesions, no erythema, uvula midline, no TTP on any teeth

CV: Hypertensive, otherwise stable

Resp: CTA B, no increased WOB

Abd: NABS, transverse healing incision s/p c-section, NT including uterus, no HSM/masses

Skin: No petechiae, no ecchymoses, no rashes, MMM, CR < 2

Ext: Edema noted on both LEs up to the knees and both hands

Neuro: CNs II-XII grossly intact, 3+ DTRs, normal motor, sensory, and cerebellar, unable to check gait secondary to patient refusal

Laboratory Studies

CBC, LFTs, renal panel, coagulation panel, and thyroid panel all WNLs

UA significant for 3+ protein, otherwise WNLs

Course

Pt was treated with Hydralazine IV x 1 and started on IV magnesium and LR in the ER, and OB/GYN was consulted. She was admitted for severe postpartum preeclampsia.

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She did well and was discharged home several days later.

Discussion

One might wonder how postpartum preeclampsia could have any relevance to pediatrics. First, it is the pediatrician, rather than the obstetrician or gynecologist, who sees the moms several days to weeks after discharge, when they bring their newborn babies in for questions/concerns, or for 2 week and later well baby checks. Since we all know that moms frequently use their child's appointment to ask questions concerning their care, it certainly is not unlikely that a mother could make comments about a bad migraine or swelling in her feet. Rather than just giving mom a prescription for Motrin, or telling her to keep her feet up, we should always remember that some of the common problems we see pre-delivery can also occur, though less commonly, postpartum. Second, this case allows some discussion of the differentials for headache, evaluation of acute hypertension, and the risk factors/consequences of preeclampsia and eclampsia. Third, adolescent patients get pregnant and have babies, and come to the pediatric clinic or adolescent clinic for their medical concerns or issues; thus, this patient might have just as easily presented to the clinic if it had been open at the time.

Headache

A common complaint in both the clinic and ER is "my head hurts." The most important immediate question to always answer first is, "Is this complaint life-threatening?" Life-threatening causes require immediate treatment and work-up, whereas non-emergent causes can be empirically treated or followed. The following is a partial list of differential diagnoses for her headaches:

1) Vascular: Carotid or vertebral artery dissection (can be secondary to trauma), subarachnoid hemorrhage (usually older patients, certainly can occur in any age; head CT if suspected with LP if negative), subdural or epidural hematoma, cavernous sinus thrombosis (secondary to infection or thrombophilia), others.

2) Infection: CNS includes meningitis, encephalitis, and brain abscess, and non-CNS includes sinusitis, OM, dental abscess or caries, herpes zoster, and other focal or systemic infections.

3) Other CNS: Tumor, pseudotumor cerebri (rare, benign, usually young, obese patient with long h/o HA, linked to OCPs, Vitamin A, and tetracycline which the patient was taking, have papilledema with normal head CT and elevated LP opening pressure).

4) Ophthalmic: Uveitis (seen in rheumatoid disorders, herpes, CMV, others), optic neuritis (multiple sclerosis, viral infections, bee stings, lead poisoning, others), and glaucoma (measure ocular pressure and > 21 is abnormal, open-angle or angle-closure types).

5) Drug-Related and Toxic/Metabolic: Nitrates and nitrites (drug of abuse, "poppers"), MAOI drugs, hypoxia or high altitude, hypercapnea, hypoglycemia, carbon monoxide poisoning, alcohol withdrawal, monosodium glutamate (used in cooking), and chronic analgesic use/abuse.

6) Miscellaneous: Malignant hypertension, preeclampsia, pheochromocytoma, fever, post-LP, TMJ disorder, and trigeminal neuralgia.

7) Primary Causes: Migraines, tension-type, and cluster headaches.

Hypertension can also have many causes, depending on whether it is sudden or chronic in onset, and how severe the hypertension is. Rather than discussing the differential for hypertension, which is very broad and beyond the scope of this

article, it is more important to discuss the management of the patient at the point in time where they have an elevated BP. First, elevated BP should always be rechecked and one must ensure the right size cuff is being used. Next, does that patient have evidence of target-organ dysfunction (encephalopathy, intracranial hemorrhage, acute LVH failure with pulmonary edema, unstable angina, acute MI, dissecting aortic aneurysm, or eclampsia); furthermore, is the hypertension new or progressive requiring treatment emergently? Although this patient did not have any of these signs of target-organ dysfunction, by definition severe preeclampsia is progressive and thus warranted immediate intervention.

Preeclampsia

Preeclampsia is a syndrome of pregnancy-induced hypertension, accompanied by proteinuria, edema, and frequently disturbances in other organ systems, that develop between the 20th week of gestation and the end of the 1st week postpartum. Hypertension is defined as either of the following: elevation of systolic BP ³ 30 or diastolic BP ³ 15 mm Hg from baseline, or elevation of BP to ³ 140/90. Proteinuria is defined as ³ 300 mg of protein in a 24-hour urine collection, or less accurately as ³ 1+ on urine dipstick. Edema is estimated by the degree of swelling or excessive weight gain; it is generally considered pathological only when it involves the face, hands and legs, and is the least reliable sign of the preeclampsia triad.

Risk factors for developing preeclampsia:

- 1) Primigravid status
- 2) Family history of preeclampsia or eclampsia
- 3) Previous preeclampsia or eclampsia

- 4) Extremes of maternal age (younger than 20 or older than 35 years old)
- 5) Preexisting hypertensive, vascular, autoimmune or renal disease
- 6) Insulin-dependent diabetes mellitus
- 7) Multiple gestation
- 8) Nonimmune or alloimmune fetal hydrops, IUGR, intrauterine fetal demise
- 9) Trisomy 13
- 10) Hydatidiform mole
- 11) Obesity
- 12) Factor V Leiden
- 13) Protein C, S, antithrombin deficiency
- 14) Antiphospholipid antibody syndrome

Criteria for Severe Preeclampsia

- 1) Blood pressure of ≥ 160 mm Hg systolic or ≥ 110 mm Hg diastolic, recorded on at least two occasions at least 6 hours apart with patient at bed rest
- 2) Proteinuria of ≥ 2 g in 24 hours or $\geq 3+$ on dipstick
- 3) Oliguria (≤ 400 ml in 24 hours) or increasing serum Cr
- 4) Cerebral visual disturbances, headache, or other cerebral signs
- 5) Epigastric or RUQ pain, nausea, or vomiting
- 6) Pulmonary edema, cyanosis, or cardiac decompensation
- 7) Impaired liver function of unclear etiology
- 8) Thrombocytopenia, hemolytic anemia, or elevated LDH, liver enzymes, or direct bilirubin (HELLP syndrome)
- 9) Fetal growth retardation

Management

The definitive treatment of preeclampsia is delivery of the fetus,

although in this particular case that treatment option was not available since it developed postpartum. Mild disease (most common form) can usually be managed conservatively, while severe disease requires treatment of the hypertension (as was done in this case with Hydralazine), prevention of convulsions (progression to eclampsia, using IV magnesium sulfate), and as mentioned previously delivery of the fetus. Although preeclampsia is seen in up to 5% of pregnancies, eclampsia is seen in less than 0.2%. Convulsions are usually preceded by headaches, epigastric pain, hyperreflexia, or hemoconcentration, but can occur suddenly in women with only mild preeclampsia.

One further point of importance is the HELLP syndrome (hemolysis, elevated LFTs, low platelets) that is a major complication that can occur in patients with preeclampsia. Although these patients appear clinically to have only mild preeclampsia, and may even be normotensive, they should be treated as severe since their risk of morbidity and mortality is profoundly elevated if prompt management is not undertaken. This should be considered in the differential diagnosis of any pregnant or postpartum patient presenting with abdominal pain.

Criteria to Establish the Diagnosis of HELLP Syndrome:

- 1) Hemolysis or abnormal peripheral blood smear
- 2) Direct bilirubin >1.2 mg/dl
- 3) LDH >600 IU/L
- 4) SGOT ≥ 72 IU/L
- 5) Thrombocytopenia (Platelet count $<100,000/mm^3$)

Complications of severe preeclampsia / HELLP syndrome / eclampsia:

- 1) Hepatic and splenic hemorrhage

- 2) End-organ failure
- 3) Placental abruption
- 4) Intracranial bleed
- 5) Death of the fetus and/or mother
- 6) Seizures

In summary, this patient had many of the clinical signs and symptoms of severe preeclampsia, and thus was at high risk of complications. She was hyperreflexic, had headache, BP $> 160/110$, and 3+ proteinuria. What complicated her diagnosis was the fact that she was postpartum. What initially we all assumed would be a migraine or other type of primary headache that we could treat with some PO or IM meds and send out quickly turned out to be something much more serious and life-threatening, and additionally was something that I and most of the other physicians I have talked with had never heard of and thus merits this discussion.

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