

PEDIATRIC NEWS

San Antonio Military Pediatric Center



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Houstaff Puzzler “Developmental Delay and Seizures”

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HPI: A 25 mo female with long history of possible seizure activity and developmental milestones presents to clinic. Pt was previously healthy until 4 months of age, when parents noticed she would occasionally bob her head up and down for a few seconds, occasionally followed by her suddenly falling to the ground. She would be limp for several seconds then recover, with normal mental status upon getting up. No history of tonic or clonic movements. No loss of bowel or bladder function. Parents report these bobbing episodes would occur hundreds of times a day, and would be increased before and after sleep.

She had normal development until around 14 months of age, when she began to lose her verbal skills to the point that she now just babbles. Pt has several odd behaviors, including tip-toeing, perseverating with objects, and hand-flapping when excited. Pt

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Eisenmenger Syndrome

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Eisenmenger syndrome ES is a topic that was probably talked about briefly when you were in medical school. It is possible, but unlikely that you have seen a patient with ES during your medical school or residency training. The logical question is: “Why am I spending time reading about this rare syndrome?”. I believe the answer is that our unique roll as military physicians puts us into environments where ES is much more common and when seeing patients internationally it is very likely that you will be asked to evaluate a patient who has ES or is on the road to develop ES if left untreated.

So what is ES? ES is a triad of progressive cyanosis, polycythemia, and eventually functional limitations that results from pulmonary obstructive vascular disease in the presence of a cardiac defect.

The Physiology

In order to understand why ES develops, let’s take a step back and look at the physiology of the cardiac lesions that if left uncorrected are typically associated the development of ES. The main lesions are atrial septal defects, ventricular septal defects, atrioventricular septal

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Scar Revision

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INTRODUCTION

The objective of a scar revision is to restore a smooth skin surface that resembles the surrounding skin. A good scar will be a fine line that is parallel to the relaxed skin tension lines. It is without abnormal pigmentation or contour irregularity. There are no contractures and no distortion of the surrounding structures. During a scar revision, steps are taken to achieve these goals.

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defects, hypertensive patent ductus arteriosus, and other more complex lesions such as double outlet right ventricle. When there is a septal defect blood has a choice in its path. The blood can either travel the direction it was originally designed to flow or it can cross the defect and take a much different path. The path the blood takes depends on the relative resistances of each path. The blood will choose the path of least resistance. This is the key to understanding how blood flows in the heart regardless what is the cardiac defect present. When you talk to a pediatric cardiologist you realize much of our language about blood flow, in, through, and out of the heart circles around the subject of resistance.

The Pathophysiology

Ventricular septal defect (VSD)

Ventricular septal defects come in many varieties most are small muscular or membranous defects that close spontaneously and never cause any hemodynamic difficulties. The defects that do cause difficulties are those that are moderate to large unrestrictive or minimally restrictive defects. The communication between the two ventricles is large enough that pressures between the ventricles essentially equilibrate. Blood enters the ventricles from the atrioventricular valves and as the ventricles contract, blood has a choice which outflow to take. As discussed above, the blood chooses the path of least resistance. In normal physiology, shortly after birth, the pulmonary vascular resistance (PVR) is lower than the systemic vascular resistance (SVR). So the blood would prefer to go to the lungs where the resistance is lower. Since this causes the pressure and the blood flow to the lungs (Qp) to be greater than the flow to

the body (Qs) and at a much greater volume and pressure than the lungs were designed to accept, cellular changes begin to transform the very nature of the lung vasculature. We usually like to repair these children prior to age one and no later than 2 years of age

Atrioventricular septal defect

Atrioventricular septal defects, AV canals, or endocardial cushion defects are really a combination of an unrestrictive VSD and primum atrial septal defect (ASD). The pathophysiology is very similar to VSDs with the exception that the volume load is usually even greater to the lungs. We usually like to repair these children at 4-6 months of age

Patent Ductus Arteriosus

A large unrestrictive patent ductus arteriosus is much less common than symptomatic ASDs or VSDs especially in the US where immunization to Rubella is common (Maternal Rubella infection is highly associated with the presence of a PDA) and few children with large ducts slip through the cracks. However, it is not uncommon to see international children with large hypertensive PDAs. These type of PDAs can cause both a volume and pressure load to the lungs and a volume load to the left side of the heart. We typically would repair these children on presentation.

Atrial septal defect

The most common defect of the atrial septum is a patent foramen ovale occurring in up to 30% of the population. However, this type of defect is rarely of any hemodynamic significance. The next most common is a secundum defect. There are also primum defects as well as sinus venosus defects. The left atrium is less compliant than the right atrium

and the end diastolic pressure of the LV is usually greater than that of the right ventricle. Therefore if there is a defect in the atrial septum blood will move from the left atrium to the right atrium across the defect. Unlike the situation with the VSDs, this is only a volume overload to the right side of the heart and lungs. It does not involve a pressure overload, seen in VSDs. This is why it is not as urgent to close large ASDs as it is to close large VSDs. These children are typically repaired around school age.

The influence of increased flow and pressure on the heart and the lungs

The common thread for all the lesions is an increased volume load to the pulmonary vasculature with or without an increased pressure load. The volume load increases the sheer stress across the endothelial cells. This starts an autocoid chain reaction that leads to an alteration of the pulmonary vasculature. In response to the abnormal physiologic state the vessels start to hypertrophy. This process takes months to years depending on the lesion and the pathophysiology. As the vasculature hypertrophies the pulmonary vascular resistance increases. In the early stages this is generally a reversible process. The problems lies when the process is no longer reversible.

Once the PVR approaches levels similar to the SVR shunting will decrease. During this time the patient who once was in failure due to overcirculation may show great clinical improvement. This is the proverbial "calm before the storm". The PVR typically continues to increase. Once the PVR is greater than the SVR then the shunts that were from the left to the right, will now be right to left. Right to left shunts cause deoxygenated blood to bypass the lungs and go back to the systemic circulation and hence hypoxia.

The Effects of Hypoxia

There are two main effects of chronic hypoxemia. These are decreased oxygen carrying capacity and shunting of systemic venous blood away from the pulmonary vasculature into the systemic vasculature. The oxygen content of the blood depends on: the hemoglobin concentration, the affinity of hemoglobin for oxygen and the partial pressure of oxygen inspired. What the body can change to increase the oxygen carrying capacity of the blood is the hemoglobin concentration. Hypoxia stimulates an increased production of erythropoietin which leads to increased hemoglobin concentration.

Additionally, in response to the hypoxia, the PVR increases and the SVR decreases. This will lead to even more right to left shunting and increased Qs because of the decreased afterload.

The Effects of Polycythemia

The increase of hemoglobin is not without its own consequences. As the hematocrit rises above 70%-75% the viscosity of the blood is greatly increased. This leads to a decrease in cardiac output and an increase in both SVR and PVR due to the increased viscosity and shear forces. Patients may complain of: headaches, general malaise, chest pain, joint pain, anorexia, dyspnea, and visual impairment. Many of these patients are also have iron deficiency anemia superimposed on their polycythemia and needs to be addressed with appropriate therapy. What has also been noted but less well understood is the apparent platelet dysfunction as well as a decreased life span of platelets.

Natural History

The natural history of ES is quite variable. Actuarial survival for

patients with ES is (1-year 77%, 2-year 69%, and 3-year 35%). Most patients survive until the third or fourth decade with the occasional rare case report of patients living into their 60's. The median age of survival for all comers with ES, including patients with ASDs who typically live much longer, is 52.6 years. Risk factors that are associated with earlier mortality are: trisomy 21, syncope, hemoptysis, lower systemic oxygen saturation, elevated right atrial pressures, SVT, earlier age at presentation, RV hypertrophy, poorer functional class, pregnancy, and VSDs, AV canals, or hypertensive PDAs.

Diagnosis

The physical findings are variable and depend on the associated cardiac lesion. Typical finds are: cyanosis at rest or with mild exertion (as the SVR drops), clubbing of the digits, right ventricular lift on examination, a loud and palpable single S2, and usually a holosystolic murmur of tricuspid regurgitation and a diastolic murmur from pulmonary insufficiency, if they are in failure then hepatosplenomegaly, peripheral edema, and ascites may also be present.

The ECG typically demonstrates right axis deviation and right ventricular hypertrophy. The chest film may demonstrate right ventricular enlargement and enlarged central pulmonary arteries. The central pulmonary arteries are typically severely enlarged with large and long standing atrial septal defects. The echocardiogram will typically show: right ventricular and right atrial enlargement, decreased RV function, pulmonary insufficiency, tricuspid regurgitation, flattening of the interventricular septum, and right to left shunting across the defect.

Cardiac catheterization is undertaken to assess the risk for surgical

repair of the cardiac defect. Patients are usually considered inoperable if the PVR is greater than 10 Wood units/m². If the PVR by 2-3 Wood units/m² after administration of 100% oxygen, nitric oxygen, prostaglandins, or adenosine, then the risk for surgery is less. Why is the risk so high if the PVR is elevated? The answer is in VSDs, hypertensive PDAs and atrioventricular canals the RV is not able to generate enough pressure to overcome the high PVR especially after cardiac-pulmonary bypass and without the help of the LV, this leads to right heart failure. As the right heart fails less blood is making its way through the pulmonary circulation and back to the LV for systemic blood flow and the cardiac output falls. As the cardiac output falls, the resulting acidosis causes the PVR to increase further causing the RV to fail more and a vicious spiral starts.

Clinical Management

The management of the patient with ES is typically not very satisfying. In fact, the author of a chapter on ES started with, "It is intellectually unsatisfying to admit that a group of patients remains for whom no 'cure' is available in modern medicine." That being said, patients with ES can be managed with treatments and medications that have shown promise for patients with primary pulmonary hypertension.

Oxygen therapy

Although not universally agreed upon, many believe that supplemental oxygen benefits patients with ES. In adults it does not appear to offer any survival advantage; but, in children supplemental oxygen during sleep appears to decrease the progression of polycythemia.

Digoxin and diuretics

Again the use of a weak inotrope like digoxin is controversial but appears to increase cardiac output. Diuretics are probably beneficial when there is significant hepatic congestion or evidence of increased intravascular volume.

Anticoagulation

It is known that patients with ES have platelet dysfunction. They are also at risk for thromboembolic events and even a small pulmonary embolus could be life threatening. Therefore, many clinicians recommend anticoagulation and aim for an INR 1.5 -2 if coumadin is used. This treatment must be balanced with the very real risk of bleeding complications such as hemoptysis.

Phlebotomy

Phlebotomy should be considered in patients whose hematocrit is > 65%, or if the patient is symptomatic at a lower hematocrit with headaches or blurry vision. The goal of phlebotomy should be to lower the hematocrit to 50-60% range and replace the volume with crystalloid or plasma. Often patients will stabilize their hematocrit levels between 60-65% and no longer need phlebotomy. Patients with ES should also be followed closely for iron deficiency anemia and be treated appropriately. Iron studies and the red cell indices should guide treatment as the absolute Hgb and HCT will be normal or high.

Vasodilator and antiproliferative therapies

For patients who respond to acute vasodilator therapies of NO or epoprostenol i.e., a >30% decrease in PVR and a similar response to sublingual therapy with calcium channel blockade are candidates for long-term calcium channel blockade.

However, where many patients with primary pulmonary hypertension respond to calcium channel blockade therapy very few ES patients will respond and in a recent study no adults with ES responded.

Another option that has had a little more success with patients with ES is continuous epoprostenol therapy. However, this is not risk free and requires a central catheter and thromboembolic events are increased.

There are some promising studies with the use of endothelin-1 inhibitors, bosentan and sitaxsentan, which have shown improvement in exercise capacity, hemodynamics, and functional class in patients with ES, but more studies are needed.

Therapeutic Options

There are basically three separate options when treating a patient with ES. 1) Symptomatic relief, 2) Surgical repair of the cardiac defect, and 3) Transplantation. Measures to be employed for symptomatic relief were described above. The management of these patients can be very challenging and humbling as what we have to offer medically is not very helpful, at this point. Attempting to correct the cardiac defect that led to this persistent pathophysiologic state is risky and it is very likely that even if the defect is successfully repaired that the pulmonary hypertension may be irreversible. The final and most severe treatment option is heart-lung or lung transplant. Patients with ES have the highest perioperative mortality of all patients and the lowest one month survival of all lung recipients. The 1-year and 5-year survival following lung transplantation for ES is 52% and 39% respectively, essentially no better or worse than without transplantation (1-year 77%, 2-year 69%, and 3-year 35%)

Restrictions

Some restrictions are prudent to improve the functional capacity and decrease the morbidity and mortality of patients with ES. Travel to high altitudes should be avoided and if a family lives at altitude then they should make every effort to move to a lower altitude, preferably sea level. Oxygen therapy on commercial airplanes is also advised to avoid a pulmonary hypertensive emergency. Pregnancy carries a very high mortality and should be avoided. Additionally, due to the increased thromboembolic risk oral contraceptives and hormone replacement should also be avoided.

Prevention

With rare exceptions, ES is a preventable disease. The key is timely repair of cardiac defects that are known to lead to ES. We are very successful at this approach in the US as access to excellent pediatric cardiothoracic surgery is not a problem. ES continues to be a significant problem in the developing world where access to surgical repair is limited if not unavailable. As military physicians we have the opportunity during humanitarian assistant missions and MEDRETEs to find some of these children before its too late for surgical repair and prevent ES. There are many non-governmental organizations whose sole purpose is to this end. Prevention is the only truly efficacious treatment option at this time.

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Role of Xylitol in Dental Decay Prevention

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Background:

Simply stated, we need four essential ingredients in order to create the “perfect storm” for dental decay (or dental caries): teeth, sugar, “bugs”, and time. Without the “bugs” (streptococcus mutans or *S. mutans*), tooth decay would not be much of a problem – even in children with high-sugar (sucrose) diets and almost non-existent oral hygiene. *The bottom line ...no S. mutans, no dental decay!*

Where does this decay-producing bacteria come from? To a large degree, tooth decay is a transmissible disease. Mothers (and to a lesser extent fathers) with *S. mutans* in their mouths are capable of inoculating their babies with these bacteria, probably through kissing and other innocent motherly (or fatherly) contact. This can occur as early as 6 months of age.

According to the National Institute of Health (NIH), nearly 20% of children between the ages of 2 and 4 have experienced some dental decay. Studies also show that children with early childhood decay

(preschoolers) are more prone for decay for the rest of their lives. By age 17, nearly four out of five adolescents have had at least one cavity. ¹ *The bottom line...in order to achieve the maximum effect, decay prevention must start early in life (before age 1)!*

Hence, today’s prevention strategies – early dental visits, fluoride application, weaning from bottles and breast, infant oral hygiene instructions are all initiated no later than the child’s first birthday.

Traditional decay prevention strategies have focused primarily on the use of two chemicals...fluoride, and to a lesser extent chlorhexidine. Fluoride is a mineral that, when applied to the tooth surface, helps harden the tooth enamel. Fluoride may either be added to municipal water supplies or it may be applied directly to teeth in the form of toothpastes, gels, rinses, and varnishes. *Fluoride has a minimal (if any) effect on bacteria!*

Chlorhexidine is an antimicrobial substance that has been used to kill oral bacteria for over 40 years. It is commonly applied directly to the tooth surface via mouth rinses, gels, or varnishes. Because of its poor taste and its propensity to stain the teeth yellow, chlorhexidine has not been popular with either adults or children.

Xylitol

A new “magic bullet” has recently surfaced in the dental prevention literature...xylitol is a natural occurring sugar, derived from fruits, vegetables, and nuts. It is classified as a sugar alcohol, or polyol. It dissolves rapidly (unlike sucrose) and has a pleasant, refreshing, cooling sensation, a clean sweet taste, with no after taste.² Unlike sucrose (a traditional sweetener produced from sugar cane and beets), xylitol has physical and chemical properties that do not

promote caries. In fact, xylitol can actually support re-mineralization (restoration of tooth enamel) where caries have already started.

Human Caries Studies

Effective caries prevention has been demonstrated with xylitol in multiple human clinical trials, predominately in Finland. These human caries studies have revealed the following:

Mothers with high *S. mutans*, who habitually consumed xylitol, showed a significant delay in the transmission of *S. mutans*.⁴

Long-term prevention – 77% caries reduction compared with controls 5 years after termination of xylitol usage.⁵

Greatest caries prevention observed on teeth that erupted after one year of xylitol usage (93% reduction) or after conclusion of the two-year program (88% reduction).³

Enhances re-mineralization by a combination of salivary stimulation and xylitol’s effect on dental plaque.⁵

Early lesions have shown partial or total radiographic disappearance with habitual use of xylitol.⁵

How does xylitol prevent dental caries?

Although xylitol and sucrose are both naturally occurring sugars, xylitol is far less cariogenic (able to promote tooth decay). Xylitol’s superiority is derived from three proposed theories:

Cariogenic bacteria process xylitol very poorly. *S. mutans* depends on sucrose and common starches in order to churn out tooth-dissolving acid. *S. mutans* ferments xylitol very poorly, which produces only very small amounts of acid.

Xylitol prevents bacteria from sticking to tooth enamel. When *S. mutans* ferments sucrose, it produces not only lactic acid, but also a

sticky polysaccharide that “glues” plaques on the tooth surface. Xylitol, unlike sucrose, does not promote plaque formation.

Xylitol promotes the colonization of less virulent strains of bacteria in the mouth. Simply put, the natural mutants selected by xylitol are generally “caries-benign” creatures that ferment slowly, do produce polysaccharides, and do not stick very well to each other or the teeth. Most importantly, their presence keeps out the more virulent strains.

What forms does xylitol come in?

The “delivery system” that produces the best anti-caries effects are those that permit xylitol to come in direct contact with the teeth for the longest time. Chewing gums that have pure xylitol candy coating work best. Brands available in the U.S. include Clen Dent, Xylichew, Xylimax and Koolerz (by Care Free).

How much xylitol is enough?

Studies show that using 4-12 grams of xylitol per day is very effective for preventing dental caries. If a piece of gum contains 1 gram of xylitol, then chewing four pieces per day would be sufficient.

How often should xylitol be used?

Clinical experience suggests that three times a day provides minimum effectiveness, while five times a day is ideal.

When should we use xylitol?

Children should start chewing xylitol gum at least one year before their permanent teeth begin erupting. To be sure, start no later than 4 to 4 ½ years of age. Studies show that teeth treated this way will be strong and have long-lasting protection.³ In fact, children who use xylitol may have just as much protection as that

provided by dental sealants.⁶

How safe is xylitol?

The amounts recommended for dental protection (up to 12 grams per day), should never cause a problem.

Who can benefit the most from xylitol?

Those at higher risk for dental decay could benefit the most from xylitol. These include, patients with:

- Rampant caries
- Poor oral hygiene
- A history of dental neglect
- A compromise of manual dexterity due to illness or injury
- Patients with orthodontic or prosthodontic appliances that tend to retain more plaque
- Athletes who consume sucrose-sweetened sports beverages

Summary

- Mothers who began chewing xylitol gum shortly after their babies were born were far less likely to transmit the cariogenic bacteria (*S. mutans*) to their newborns in the first 12 years of life.^{4,5}
- If children get in the habit of chewing xylitol gum (or using other xylitol products) at least one year before the eruption of their permanent teeth, they will likely be protected from tooth decay for the rest of their lives.³

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For a complete list of all xylitol publications since 1971, go to www.xylitol.org.



New Roads in the Management of Rhinitis

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The treatment of rhinitis is evolving in parallel with increased understanding of nasal physiology. Newer technologies combined with immunology

research is allowing us to have a better understanding of nasal function. Concepts of upper airway inflammation, mast cell activation, early and late-phase reactions, and cytokine function are now being applied to pharmaceutical research. This demands a new look of our approach to rhinitis and its therapy.

In allergic disorders, histamine is an important mediator of inflammation. Older and newer antihistaminics are the most diverse class of medications used to relieve symptoms of rhinitis. The first generation (older antihistaminics) medications have a variety of side effects to include impairment of psychomotor performance, sedation, dry mouth, urinary dysfunction and, in high doses, may produce cardiac toxicity.

The newer second generation H1 antihistaminics such as cetirizine, fexofenadine and loratadine have a more favorable benefit-to-risk ratio than its predecessors. They have excellent H1-blocking properties in the airways and skin, as well as having anti-allergic and anti-inflammatory activities. They also have optimal pharmacokinetic and pharmacodynamic characteristics in clinical pharmacology studies in humans. These characteristics include rapid absorption after oral administration, absence of interaction with concomitantly administered medications, rapid onset of action, 24 hour duration of action after a single dose, and absence of tachyphylaxis. Most of them consistently relieve symptoms of itching, sneezing and rhinorrhea, and improve quality of life in patients of all ages.

In comparison to first generation H1 antihistaminics, the second generation medications are much less likely to cross the blood-brain barrier, impair psychomotor performance or cause sedation. Second generation H1 antihistaminics do not

cause dry mouth or urinary dysfunction. In addition, these medications are free from cardiac toxicity at the recommended FDA dosing and even if taken in overdose.

A newer generation of antihistaminics are currently being studied (levocetirizine, norastemizole) or are currently in the market (desloratadine-Clarinet). These newer group will have less side effects and will be more effective in controlling allergic disorders.

We await for new surprises in the near future. Newer drugs will emerge and will have an important role in the management of patients with allergic disorders. Gene Therapy, Cytokine Therapy, and anti-IgE Therapy is currently under investigation and the response to treat allergic disorders is very promising. Ultimately, better therapeutic options should decrease symptoms and improve quality of life, reverse pathogenic and physiologic changes, have low side effects, and be cost effective.



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INDICATIONS

Scar revision may be requested for several reasons. Scars may be revised because of aesthetic concerns, instability, functional impairment, or pain. The scar must be evaluated relative to the patient's concerns and expectations. Unrealistic expectations are the most common cause of failure in scar revision. Patients and family must understand that all wounds will result in scarring of the tissues. Scar revision is directed at creating an

optimal scar, not eliminating it completely

HISTORY

The patient evaluation can be divided into three parts: (1) the history, (2) the examination, and (3) the planned treatment. The history can help to determine why the scar is unacceptable. Something must be improved if the result is to be better than the previous repair. The etiology of the scar deformity can be related to factors attributed to the patient, the wound, or the management.

PATIENT FACTORS

Some patients are prone to developing unsightly scars. Scars tend to be more visible in dark skinned individuals. They more frequently develop keloid scars than light skinned people. Some genetic abnormalities, such as Ehlers-Danlos syndrome, cause patients to develop abnormal scars. In general, an older patient will have a better scar than a younger one. Patients aged 2-20 years old tend to have a longer period of scar maturation, so they must be cautioned that their scars will be red and raised for a longer period of time.

WOUND FACTORS

Some areas of the body are prone to developing unsightly scars. Scars located in the triangle formed by the xiphoid process and tip of the shoulders tend to become wide and thickened. The skin tension caused by the pull of the chest muscles and upper extremities result in a widened scar. Other troublesome areas are the shoulders, knees, and lateral submandibular area. The tension forces across the joint will cause the healing scar to widen. Tissue loss at the time of the trauma will lead to wound tension, which tends to result in a larger scar. Scars with

particulate material embedded in the tissue will be prominent because the particles tattoo the skin. The contractive forces on circular shaped wounds cause the central area to become prominent, creating a trapdoor deformity.

INITIAL WOUND MANAGEMENT

A history of the initial wound management is required to find areas to improve during scar revision. Poor technique, infection, wound dehiscence, and secondary healing can lead to unacceptable scars. Improvements must be made if the outcome is to be better with scar revision. If the treatment and post-operative course were optimal, subsequent attempts at scar revision are unlikely to result in improvement.

THE AGE OF SCAR

The age of the scar determines the optimal time for scar revision. Scars will tend to improve with time as the scar matures. The cellular matrix of the mature scar is different from an immature one. As the scar matures, hyaluronic acid is replaced by chondroitin-4-sulfate and collagen cross-linking increases. The mature scar is flat, without redness, and no longer itches. The classic period of scar maturation is 12 months, but can be shorter or longer depending on the age of the patient. Scars may mature earlier in elderly patients. Scars in children may require up to 48 months to mature. If the scar is allowed to mature, scar revision may become unnecessary. Scars that do not have targets for improvement should be allowed to mature prior to attempting scar revision. But, when there are obvious technical deficits, it is not necessary to wait a specified period before revising the scar.

CONTRAINDICATIONS BASED ON HISTORY

Scars in children have a prolonged maturation phase. Scars also tend to widen with body growth. For these reasons, optimally treated scars should not be revised until the child reaches maturity (14 to 16 years in girls, and 15 to 17 years in boys). Local or systemic diseases contributing to the poor scar outcome must be controlled prior to scar revision. Patients with unrealistic expectations of the scar revision should also be avoided.

EXAMINATION

SCAR LOCATION

Scars are visible because of color differences between the scar and the surrounding skin, because of shadowing caused by contour differences, or misalignment of anatomic structures. Immature, red scars will show up against the surrounding tissues. The raised nature of the scar will be visible as light is reflected from the surface. Similarly, tattooing of debris and road dirt will cause some traumatic scars to be prominent. Scars that have even 2 mm of misalignment of the vermilion border of the lip will be visible from several feet away.

PHYSICAL CHARACTERISTICS

Scars contract along their length. Long scars across flexion creases can bowstring as they contract. Interruption of the scar into smaller scars may improve function. Wounds allowed to heal secondarily result in wide scars.

RELAXED SKIN TENSION LINES (fig 1)

The relaxed skin tension lines can be found by pinching the skin to relax it. The skin will easily form folds and wrinkles in one direction but not in the direction perpendicular. The relaxed skin tension lines coincide with facial wrinkle lines in



Figure 1

most instances.

Incisions placed parallel to the relaxed skin tension lines result in superior scars. The maximal skin laxity is perpendicular to the relaxed skin tension line and aligned with the direction of the muscle pull. The incisions placed parallel to the skin tension lines will be pulled closed by the skin tension. On the other hand, incisions at right angles to the skin tension lines are pulled apart resulting in a widened scar.

In the forehead, the frontalis muscle pulls vertically. The relaxed skin tension lines and desired scar direction is horizontal. A vertical scar will be pulled wider by the skin tension whereas a horizontal scar is pulled closed by the skin tension.

CONTRAINDICATIONS BASED ON EXAMINATION

Findings that preclude scar revision include immature scars, recurrent scars in unfavorable locations, recurrent keloids and hypertrophic scars previously treated optimally. Local skin conditions such as acne, or systemic disease must be corrected prior to surgery.

METHODS OF SCAR REVISION

FUSIFORM SCAR REVISION (fig 2)

Fusiform scar revision is the most common method of scar revision. It is most useful for narrowing widened scars. This

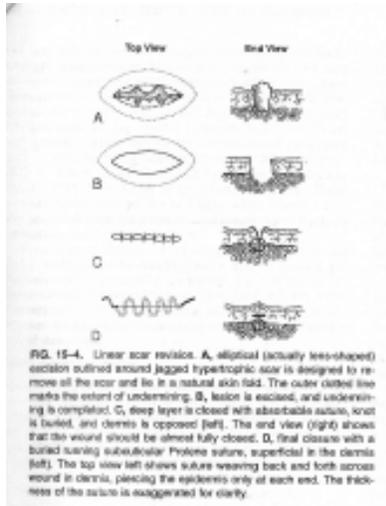


Figure 2

method can also be used to line up anatomic landmarks, such as the eyebrow. The scar is excised in a fusiform shaped portion of tissue. The skin edges are then directly approximated. The long axis of the fusiform incision should be parallel to the relaxed skin tension lines to obtain an optimal scar. Reducing tension on the healing scar will result in a finer scar. The scar can also be designed so it will lie within natural wrinkle lines or boundaries.

Z PLASTY (fig 3)

A Z-plasty uses a Z-shaped incision and transposes triangular flaps. It can improve a scar by

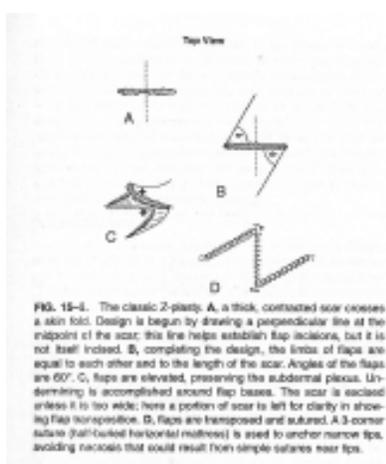


Figure 3

breaking it into smaller portions and changing the direction of the scar to lie parallel to the relaxed skin tension lines. The Z-plasty procedure changes the direction of the central limb following transposition. It also results in an increase in scar length that varies with the angles of the Z. Angles of 30 degrees increase the length by 25%, angles of 60 degrees increase the length by 75%, and angles of 90 degrees increase the length by 125%. The scar length increase is useful in correcting scar contractures. The Z-plasty is useful for correcting burn scar contractures of the upper extremities.

W PLASTY (fig 4)

The W-plasty is useful for breaking up straight-line scars, but without the tissue movement required by a Z-plasty. It is created by a series of small incisions linked at sharp angles to create the appearance of multiple Ws linked together. It can be used to camouflage long scars on the face that do not follow the relaxed skin tension lines.

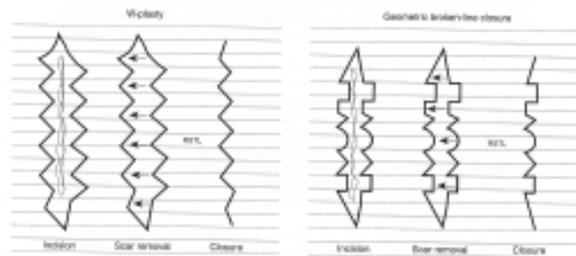


Figure 4

GEOMETRIC BROKEN LINE CLOSURE (fig 4)

The geometric broken line closure is used to camouflage scars by making the scar irregular. The existing scar is excised in a series of squares and triangles. The eye does not readily follow the haphazard scar making it less visible.

DERMABRASION

In dermabrasion, the superficial

layers of the skin are removed using a rapidly rotating wheel. It can be a wire or a fine grinding wheel, and is used to plane the skin down to the interface between the papillary and reticular dermis. Dermabrasion is used to level the scar with the surrounding tissues, usually by removing some of the surrounding normal skin. It is useful for treating depressed acne scars as the leveling of the skin surface makes the shadowing less prominent.

KELOIDS AND HYPERTROPHIC SCARS

Keloids and hypertrophic scars can both present as unacceptable scars and it may be difficult to distinguish between them. Both can present as thickened, elevated scars. The distinction is important

because the natural history and treatment options are different.

Keloids extend beyond the borders of the original wound, whereas hypertrophic scars usually remain within the confines of the original injury. Most hypertrophic scars will flatten and fade as they mature, but keloids will often continue to grow. Hypertrophic scars are frequently seen in areas where the wound healed under tension. They can occur at any age and any part of the body. On the other hand, keloid formation is seen more frequently in younger patients. They appear to be genetically related and are more frequent in dark skinned people. There appears to be

a correlation between melanocyte stimulating hormone and the incidence of keloid formation.

The prognosis is better for hypertrophic scars because they tend to flatten and blend with time. Scar revision can be beneficial if tension is avoided on the healing wound. On the other hand, keloids tend to recur following excision. Excision alone results in a 45 – 100% recurrence rate for keloids.

Pressure has been used in the management of burn scars since the 1970s. It can be used to manage hypertrophic scars. Pressure applied over prolonged periods is thought to decrease tissue metabolism and increase collagenase activity. Pressure causes the collagen bundles to be reoriented resulting in less thickness to the scar. The recommended pressure is 24 – 30 mmHg and used continuously for 6 - 12 months.

Silicone gel is used in the treatment of thickened scars. Although its mechanism is unclear, and its use was initially met with skepticism, there is now good evidence for its efficacy in treating both hypertrophic scars and keloids. The gel is worn continuously over the scar. It is more useful for treating hypertrophic scars versus keloids.

Steroids are often used pre and post-operatively to decrease recurrence. Surgery combined with steroid injection reduces the keloid recurrence rate to less than 50%. Triamcinolone acetate (kenalog) can be injected directly into the scar to induce regression. Intralesional steroid injections are painful. Steroids that spread into the surrounding tissues can cause skin atrophy, telangiectasias, and hypo pigmentation.

An alternate treatment is radiation therapy. Radiation induces micro vascular changes in the tissue that decrease collagen synthesis. Recurrence following surgery with postoperative radiation is approximately 25%. When surgery, radia-

tion, and steroid injections are combined, the recurrence rate is reduced to 10%.

Lasers have been used to treat keloids and hypertrophic scars. The yttrium-aluminum-garnet (YAG) and pulsed dye lasers are used to selectively ablate blood vessels and decrease erythema. Unfortunately, its efficacy in treating hypertrophic scars was not demonstrated in a randomized, controlled study. Laser therapy remains emerging technology with promise, but still requiring further study.

Adhesive microporous paper tape is felt to be beneficial to scars. Its effect is thought to be part mechanical from the pressure, and part occlusive, similar to silicone gel therapy. It can be used as a preventative measure in patients. The tape can be easier to maintain over mobile areas such as joint surfaces.

Cryotherapy is beneficial for treating keloid scars. It can result in flattening in 51 – 74% of patients. Its use is limited by delayed healing following treatments. Cryotherapy also can result in pigment abnormalities, skin atrophy, and pain. It is generally used only for very small scars.

There are several emerging therapies with promise in the future. Interferon, intralesional 5-fluorouracil, and bleomycin injections appear to be useful for treating problem scars. Interferon has been shown to increase collagen breakdown. However, the pain associated with injection is significant. Similarly, intralesional 5-fluorouracil has been shown to be useful for difficult to treat patients. Bleomycin can be useful for patients who do not react to steroids, but its use in wart treatment is reported to cause nail loss and Raynaud's phenomenon. Larger scale studies are warranted with these treatments prior to considering them standard treatments.

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Continued from page 1

has also regressed in her gross motor skills, and has recently started crawling instead of walking. She has also had arrest of OFC growth for the past 4-5 months.

PMHx: Negative, No hospitalizations, uncomplicated term pregnancy.

FHx: Uncle with seizure disorder

Physical Exam: Wt 14.8 kg (90%), Length 88 cm (50-75%), OFC 48 cm (50%) BP 122/71, HR 122, RR 28, T 97.3, O2 Sats 99% on RA

General – Well nourished, no acute distress, interacts poorly with examiner, babbling, no words heard.

HEENT- deep-set eyes, frontal bossing. TMs clear, OP clear, supple neck, nl fundoscopic exam.

Neuro – Poor interaction with examiner. EOMI, PERRLA, CNs II-XII grossly intact, mild decrease in tone throughout, nl strength, nl sensation, +2 DTRs, Babinski with downward toes, Gait much more clumsy than expected for age.

Skin – no lesions; exam with wood’s lamp negative.

Remainder of the exam was unremarkable.

Hospital Course: Patient was admitted to facilitate workup. Video EEG showed myoclonic seizure activity concerning for a progressive myoclonic epilepsy. Head CT and MRI were normal. All labs were unremarkable, which included transferrin, serum & CSF lactate, thyroid function tests, biotinidase, pyruvate, ammonia, and serum amino acids. Patient was discharged home with suspicion for Rett Syndrome, and the test for a mutation in the

MECP2 gene is pending.

Discussion

Rett Syndrome is a syndrome affecting young girls characterized by a period of apparently normal development for the first 6-18 months of life, followed by profound cognitive impairment, communication dysfunction, stereotypic movements, and pervasive growth failure¹. The incidence in Texas is 1:22,800, and diagnosis is largely based upon clinical characteristics².

Clinically, the syndrome is remarkable for apparently normal prenatal and perinatal periods, normal psychomotor development until around 6 months of age, deceleration of head growth starting around 3 months, decrease of purposeful hand skill starting around 6 months, communication dysfunction and social withdrawal starting around 9 months, stereotypic movements starting at around 1 year of age, impaired or absent ambulation, and absence of other disease processes (no organomegaly, optic atrophy, retinal changes, or history of IUGR). Stereotypic movements often consist of hand wringing, hand clapping/patting, and hand mouthing. Definitive diagnosis is made by identification of a mutation in the MECP2 gene, which encodes methyl-cytosine-guanosine (CpG) binding protein 2 (MECP2).⁽³⁻⁴⁾

Patients typically survive into middle age, with 70% living to 35 years old. Patients often have a mental age at the 8-10 month level and gross motor function at the 12-18 month level. Most adaptive skills, such as feeding and dressing, are

never acquired effectively. There is a large range in the reported seizure incidence, from 30% to 80%. EEGs are almost always abnormal after the age of 2 years, making it often difficult to differentiate behavioral patterns from seizures. Only about 60% of girls with Rett Syndrome remain ambulatory. The growth failure is pervasive, often falling off the growth curve, though many reach the 5th percentile again at around 7 years of age.

The MECP2 gene is transmitted in an X-linked dominant pattern, with presumed lethality in males. More than 99% of cases in women are sporadic mutations. The gene mutation for MECP2 is identified in 80-85% of females with classic Rett Syndrome, and in less than 50% of those diagnosed with atypical Rett Syndrome.

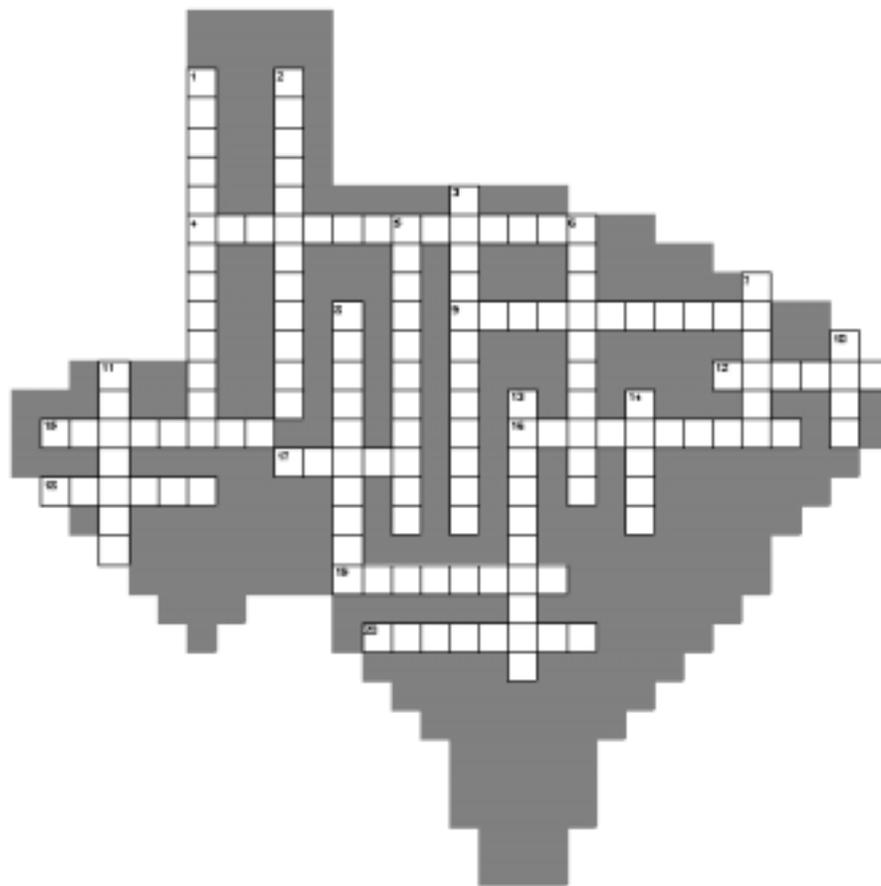
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The information and opinions stated in the *Pediatric News* are the opinions of the authors and in no way reflect official policy or medical opinion of the United States Army or any other government agency.

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Pediatric News Review



Word List

- actionplan
- asthma
- botox
- cariogenic
- chlorhexadine
- chorea
- controllers
- delivery
- fluoride
- guidelines
- hydronephrosis
- left
- menstruation
- mitral
- preeclampsia
- pyeloplasty
- seizures
- spirometry
- Wilms
- xylitol

Across

- 4. 60% of all neonatal occurrences of this will be due to UPJ obstruction
- 9. Medications used on a daily basis to treat persistent asthma
- 12. Irregular, spasmodic, involuntary movements of the limbs or facial muscles, often accompanied by hypotonia
- 15. Most effective treatment of preeclampsia
- 16. An important written piece of asthma education
- 17. The most common oncologic cause of hematuria and an abdominal mass is due to this tumor
- 18. bicuspid valve between the left atrium and ventricle, named for its resemblance to a Bishop's miter
- 19. Classic symptom of eclampsia
- 20. Mineral that helps strengthen the tooth enamel

Down

- 1. Antimicrobial substance that kills oral bacteria
- 2. A common cause of factitious hematuria in an older female child
- 3. Triad of hypertension, edema and proteinuria
- 5. The surgical procedure for open correction of UPJ obstruction is called _____
- 6. The best objective test of airways obstruction
- 7. The most common chronic illness in children
- 8. Instruments that help standardize disease management using evidence based medicine
- 10. The majority of all UPJ obstruction is located on the _____ side
- 11. Natural occurring sugar alcohol derived from fruits, vegetables and nuts
- 13. Able to promote tooth decay
- 14. Competitive inhibitor of acetylcholinesterase at post-synaptic motor endplate