PEDIATRIC NEWS

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R.B.- Housestaff Puzzler

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Patient is a 15-year-old female who presented to the adolescent clinic for follow up of her ulcerative colitis and for coordination of her medical care. Approximately five years ago, she was hospitalized for evaluation due to three months of intermittent migratory arthralgias and joint effusions of bilateral knees, hips, ankles, and left wrist. She also had multiple episodes of spiking fevers and the day prior to admission had 4 episodes of bright red blood per rectum. A colonoscopy was performed which revealed active inflammation to the hepatic flexure and the biopsy showed acute colitis with individual necrosis. The EGD and bone marrow were normal. An ESR at that time was 82, rheumatoid factor 80, ANA was negative, LFT normal. Patient had a normochromic, normocytic anemia. Patient was diagnosed with ulcerative colitis and treated inpatient with solumedrol and sent home on prednisone. As an outpatient, she saw ophthalmology with a normal exam. Since diagnosis, civilian providers have treated her for

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"You Are What You Eat!"

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When "The Band" recorded the John Simon / Peter Yarrow song "You Are What You Eat," I had no idea how prophetic they were. Over the past 23 years nutrition has been an important part of the work I do. In this article I would like to discuss how what you eat affects your athletic potential, your neurologic capabilities, and your risk for disease and injury.

Specifically I would like to discuss the role of 3 omega and 6 omega fatty acid intake. In the scientific literature, you may see these written as 3 omega and 6 omega or n-3, n-6 fatty acids. They may also be called 3 omega and 6 omega polyunsaturated fatty acids or PUFA. Whichever name is applied these are the same compounds. I am sure some, if not all of you are saying to yourselves, "What on earth is he talking about?"

Every cell in our body has a wall called the cell membrane that surrounds it and shapes the cell. This membrane is a living interface that directly affects how the cell functions and how it is viewed by our immune system. Each of us strives to maintain our health by aerobic and anaerobic exercise, eating healthy, while maintaining flexibility and agility through stretching exercises. Every cell in our body is striving to maintain that same objective: flexibility, agility and adaptability. For our cells this comes as fluidity and compressibility, and is accomplished by the many membrane functions.

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The Pediatric Management of the Parent with Borderline Personality Disorder

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Background

An illness in a child constitutes a time of great uncertainty and stress for families. When the parent has borderline personality disorder (BPD), the situation is often catastrophic for the parent, the child, and the medical staff. You should consider this

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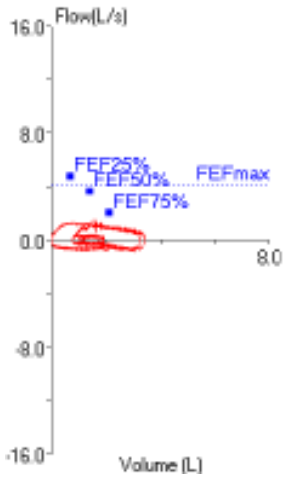
ulcerative colitis with sulfsalazine and folate.

Patient developed recurrent sinusitis and epistaxis with first episode per chart approximately three years prior to presentation in the adolescent clinic. She had been treated with multiple courses of two to four weeks of antibiotics without improvement. A CT showed erosive sinusitis so patient was taken to surgery. During surgery, patient was intubated with a 6.5 mm cuffed endotrachial tube. No organisms were isolated from sinus biopsies. One month later she was still symptomatic and was taken back to the operating room. Of note, during the second surgery she was unable to be intubated with the same size endotrachial tube and required a 5.0 uncuffed tube. It was after this second surgery that patient presented to the adolescent clinic for evaluation.

In the clinic, patient reported a slight unintentional weight loss of 6 pounds from her baseline104 pounds. Vital signs were all within normal limits with a respiratory rate of 16 and oxygen saturation of 100% on room air. Physical exam was normal except for audible low-pitched inspiratory and expiratory stridor at rest without retractions or flaring and erythematous nasal mucosa. A lateral neck x-ray was

obtained which revealed significant subglottic stenosis. She had multiple labs drawn and was sent to pulmonology for evaluation of her airway. Spirometry revealed complete flattening of both inspiratory and expiratory loops consistent with severe subglottic stenosis.



Fiberoptic bronchoscopy revealed only active inflammation without evidence of chronic scarring. A neck CT with 3-D reconstruction showed the subglottic lumen narrowed to 4.8 millimeters with a normal trachea. The chest CT was without evidence of pulmonary involvement.

Multiple laboratory tests were ordered. A complete blood count was normal with a hematocrit of 38, platelets 396. ESR and CRP were significantly elevated at 91 and 1.2 respectively. Chemistry was normal with a BUN of 8 and creatinine of 0.6. The urinalysis was also normal with less than 1 red blood cell per high-powered field and no protein. Rheumatoid factor was elevated at 25.9. P-ANCA, ANA panel were negative. Immunoglobulins were normal except for IGG elevated at 2088. A review of the colonoscopy done five years ago called the diagnosis of UC into question as only active inflammation was observed without evidence of chronic disease. The diagnosis was obtained when the proteinase 3 (C-ANCA) came back significant for disease at 1.66 with the upper limits of normal being less than 0.8. Patient was diagnosed with Wegener's Granulomatosis and admitted for three days of IV Solumedrol 1 gram/m2 as well as consults to rheumatology, ENT, pulmonology, and nephrology. She was then placed on prednisone 1 mg/kg by mouth every day and methotrexate 10 mg/m2 subcutaneously once a week for 2 months with the plan to reevaluate in 2 months. Folic acid was started due to the methotrexate and she was placed on bactrim for PCP prophylaxis. Several weeks after diagnosis, a balloon dilation of the subglottic space with topical mitomicin-C placement to minimize scarring was performed. A biopsy was obtained at this time to confirm the diagnosis. She does not currently have any evidence of renal or pulmonary involvement.

Wegener's Granulomatosis

Vasculitides are defined by the presence of leukocytes in the vessel wall with reactive damage to the vessel that can lead to ischemia of the tissues and necrosis. Wegener's Granulomatosis is a systemic vasculitis of both the medium and small arteries and may occasionally involve the large arteries. It is rare in children occurring in less than one per two million with the true incidence unknown. The male to female ratio is reported as1:1; however, one study examining differences in 28 patients diagnosed with Wegener's in Switzerland between 1991 and 1998 found 20 cases to be female suggesting a female predominance in childhood. The mean age of presentation is 40 years although fifteen percent of patients present prior to 19 years old. Two forms exist, the classic form and the limited form of the disease. The classic form

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involves the upper and lower respiratory tract as well as the kidneys while the limited form tends to involve the upper respiratory tract or the lungs. Although the limited form is found in approximately 25 % with the disease, 80% ultimately develop renal involvement so it is all likely a spectrum of the same disease.

Multiple organs may be involved with this disease to include the joints, eyes, skin, nervous system, heart, gastrointestinal tract, subglottis or trachea, lower genitourinary tract, parotid glands, thyroid gland, liver, and breast. Joint involvement may present as effusions, arthralgias, or arthritis. Eye involvement may present as conjunctivitis, corneal ulceration, episcleritis, optic neuropathy, nasolacrimal duct obstruction, proptosis, diplopia, retinal vasculitis, and uveitis. Vesicular, palpable purpura, ulcers, subcutaneous nodules, or hemorrhagic lesions may appear on the skin. Patients with Wegener's granulomatosis may present with nervous system abnormalities such as mononeuritis multiplex, cranial nerve abnormalities, CNS masses, external ophthalmoplegia, or hearing loss. Pericarditis, myocarditis, and conduction system abnormalities have also been noted in patients with this disease.

Patient most commonly present with persistent rhinorrhea, purulent or bloody nasal discharge, oral and/ or nasal ulcers, polyarthralgias, myalgias, or sinus pain. Other presenting symptoms include cough, dyspnea, hemoptysis and pleuritic chest pain which may be accompanied by pulmonary consolidation, alveolar opacities associated with alveolar hemorrhage, nodules ranging from a few mm to 9 cm which often cavitate, or pleural opacities on chest x-ray. Nonspecific complaints include fever, night sweats, anorexia, weight loss, malaise. Renal involvement usually involves microscopic hematuria with or without red cell casts and variable degrees of renal insufficiency. It may rarely present with extra thoracic masses outside the lung with the most common locations involving the breast and kidney. Childhood onset Wegener's Granulomatosis presents fives times more frequently with subglottic stenosis than adult onset with the other aspects of the disease occurring with a similar frequency.

Since 1985 when the presence of ANCA was first associated with Wegener's Granulomatosis, its diagnosis has been suggested from the presence of circulating antineutrophil cytoplasmic antibodies directed against proteinase 3 (C-ANCA). Proteinase 3 is the target antigen of these antibodies and is located in the azurophilic granules of neutrophils and the peroxidase-positive lysosomes of monocytes. It must be differentiated from P-ANCA that is directed against myeloperoxidase found in another ANCA+ disease, microscopic polyangiitis (MPA). Using immunoflouescence testing of serum using ethanol-fixed neutrophils as the substrate, sera containing anti-proteinase 3 antibodies demonstrate a cytoplasmic pattern of staining (C-ANCA) while myeloperoxidase antibodies produce a perinuclear pattern (P-ANCA). ANCA is found in approximately 90% of patients with active Wegener's Granulomatosis, therefore, a small subset is present that are not ANCA+. However, although C-ANCA appears to be specific for Wegener's with 80-95% with ANCA being C-ANCA+, the sensitivity of it may be low in the range of 65 to 70% of patients with the limited form of the disease. However, distinction between MPA and Wegener's Granulomatosis is not clinically important as treatment and prognosis is identical in both disorders. Also of note ANCA, especially P-ANCA, has been reported in virtually all inflammatory rheumatologic conditions so all information must be taken into consideration prior to making a diagnosis.

Common laboratory abnormalities include leukocytosis, thrombocytosis greater than 400,000/mm3, elevated ESR, and a normochromic, normocytic anemia. The diagnosis is normally confirmed by tissue biopsy at a site of active disease for two reasons: first, C-ANCA may not be sufficient to give the diagnosis and second, treatment is toxic. Biopsy frequently reveals granulomatous inflammation; occasionally the actual vasculitis may be observed in 1/3 of biopsies. Although biopsy of the upper respiratory tract is less invasive, renal or lung biopsy is possible. Renal biopsy typically reveals a segmental necrotizing glomerulonephritis with little or no immunoglobulin deposition called pauci-immune on immunoflouescence or electron microscopy. The usual histopathologic finding on lung biopsy is vasculitis and granulomatous inflammation.

Untreated patients die within two years due to respiratory or renal failure. Treatment favored by most physicians includes daily oral cyclophosphamide (1.5 to 2 mg/kg per day) and corticosteroid (1 mg/kg prednisone per day for one month followed by prolonged taper) combination therapy to induce remission that is usually achieved in months to two years. If patients present with fulminant renal or respiratory disease, initiation of therapy is different with 250 to 1000 mg IV solumedrol given the first three days followed by the prednisone and a higher dose of oral cyclophosphamide (3-4 mg/kg per day). Remission with this regimen

has been found by several studies to be in the range of 85-90% with complete remission obtained in 75%.

After remission, several regimens substitute less toxic and immunosuppressive therapy for the cyclophosphamide. Cyclophosphamide therapy has been associated with amenorrhea greater than one year(57%) or infertility, cystitis (50%), bladder cancer (5.6%), myelodysplasia (2%), lymphoma (0.7%). Corticosteroid use was associated with an increased risk of opportunistic infections, cataracts (21%), diabetes mellitus (8%), osteopenia, fractures (11%), and aseptic necrosis of bone. Ninety percent of patients experience persistent morbidity from this disease despite adequate treatment to include hearing loss (35%), cosmetic and functional and nasal deformities (28%), tracheal stenosis (13%), and permanent renal insufficiency (42%) with segmental sclerosis being most common finding on biopsy. Bactrim prophylaxis has decreased pneumocystis carinii pneumonia in these patients which had been a deadly complication affecting approximately 6% of patients on this regimen.

Alternative treatments are many include monthly IV cyclophosphamide, methotrexate, corticosteroids alone, and plasmapheresis. Many drugs have been tried and are beyond the scope of this article. However, given the fact that our patient is considering alternative therapy given the toxicity of daily oral cyclophosphamide, her therapy will be briefly discussed. Monthly IV cyclophosphamide, in an effort to decrease the cumulative dose, has been found to produce variable results currently attributed to differences in the disease severity. Methotrexate with prednisone may be helpful in patients with active but not life-threatening disease. Both corticosteroids alone and plasmapheresis have not been found to be an effective alternative. Plasmapheresis has been found to be a useful adjunct in patients with concurrent anti-glomerular basement membrane disease, those with dialysis-dependent renal failure, and those with severe pulmonary hemorrhage.

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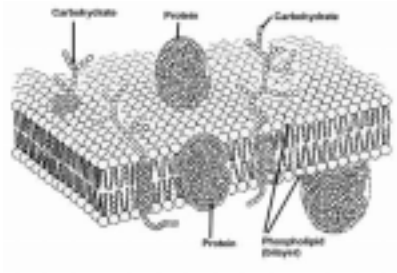
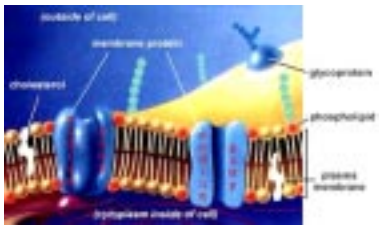
The cell membranes are composed of fat molecules arranged into a lipid bi-layer. The illustration below shows this phospholipid bilayer.



This is where our intake of fatty acids in the form of 3 omega and 6 omega fatty acids becomes important. We humans can not manufacture either 3 omega or 6 omega fatty acids, yet they are essential for the formation of every cell in our body. This is why they are called essential fatty acids. Our nerve cells conduct signals and release neurotransmitters based upon the stability of the nerve cell membranes. The method by which a nerve signal travels down a nerve is called depolarization. The method by which a nerve cell depolarizes is by conducting an electrical charge across the cell membrane by moving sodium molecules. The sodium moves through a sodium channel. These sodium channels are always located in the cell membrane adjacent to a 3 omega phospholipids. Therefore our nerves will transmit these electrical signals better if there is approximately one 3 omega fatty acid for every two to four 6 omega fatty acids. In the illustration below the red phospholipids are 3 omega and



the yellow are 6 omega. (Sorry, no color)

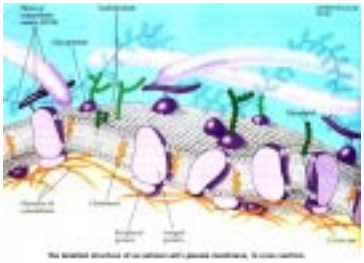
In addition to transmitting signals in a more appropriate fashion, our intake of 3 omega and 6 omega fatty acids affects the synthesis of cellular proteins and enzymes which affect the release of neurotransmitters, and the recovery of the retinal rods and cones we use for vision. These discoveries are the reasons many infant formula companies have added the 3 omega fatty acids DHA and EPA to their formulas (Similac® Advance® and Enfamil® Lipil®). Several studies have indicated improved visual and cognitive development when these phospholipids are supplied in amounts similar to those in breast milk and in the 1:3 ratio found in breast milk.

In recent years, the capability to measure the phospholipid ratios in red blood cell membranes has suggested that several neurologic conditions, which have been increasing in incidence throughout the industrialized world, may be related to relative deficiencies of 3 omega fatty acids. Longitudinal studies in several industrialized countries have shown an increase in several diseases that correlates with a declining prevalence of 3 omega fats, and an increasing presence of 6 omega fat in the diet. These diseases include: Depression, Attention Deficit Disorder with and without Hyperactivity, Alzheimer Disease and Dyslexia. By changing the ratio of 3-6 omega fats C in our diet, we are changing the velocity at which nerves transmit signals, and reducing the amount of neurotransmitters released at the synapses. This has upset the balance in patients that may be genetically predisposed to these disorders and increased the incidence of these illnesses in industrialized nations.

For those who wish to pursue more in-depth information, a list of references, which you can have your library obtain, is included at the end of the article. Hopefully, I am convincing you that the ratio of the fat we eat is critically important to our functioning at peak capability from a neurologic perspective. Unfortunately for those of us who have followed current dietary trends, neurologic performance below our peak capabilities is not the only harm that may befall a diet deficient in 3 omega phospholipids.

Our immune system as well as our nervous system evolved over 10,000 years ago. Our diet has changed with the agricultural and industrial revolutions, with the most drastic changes occurring over the past 50 years. For the previous 10,000 years our diet consisted of a ratio of one 3 omega acid for every two to four 6 omega acids. This reflected a diet that was primarily vegetarian with lean meats, fish, green leafy vegetables, fruits, nuts, berries and honey. Cereal grains have largely replaced the leafy vegetables, fruits, nuts and berries in our diet, and beef has largely replaced fish and lean meats. Currently most Americans are consuming a ratio of 16 (6 omega) to each (3 omega) fatty acid. Wheat, maize, and rice make up 75% of the world's grain production; they contain only 6 omega fatty acids. This disproportionate intake of 6 omega fats adversely affects the function of our immune system.

The effect of excess 6 omega fats on immune function occurs for several reasons. The 6 omega and 3 omega fats differ in the carbohydrates and glycoproteins that attach to them along the cell surface. Some of these proteins are HLA antigens. These can either be antigens that tell immune cells that all is well in the neighborhood (down regulatory) or they may warn the immune system that this cell is sick or stimulate an immune response (up regulatory).



It's much like a nice subdivision. If homes are situated with a few trees surrounding them, the community appears more attractive when we drive by. However, if there are no trees around the homes, it is not as appealing to the eye. The immune system evolved recognizing neighborhoods with trees as normal and those with few trees as abnormal.

By eating a diet deficient in 3 omega we can increase our likelihood of developing autoimmune diseases or inflammatory conditions. These include autoimmune thyroiditis, autoimmune diabetes, allergies, asthma, arthritis, atherosclerotic heart disease, inflammatory bowel disease, and cartilage degeneration. These diseases occur because the cell membranes do not appear normal to the immune system and a destructive process ensues. In addition, when we consume a high intake of 6 omega fats we up regulate the lipoxygenase pathway and promote the production of pro-inflammatory cytokines. These are molecules in our body that encourage our immune system to be more aggressive. Medications such as aspirin, indomethacin, ibuprofen, and prednisone are designed to reduce the production of these lipoxygenase produced cytokines. In contrast, the 3 omega fats promote the cyclooxygenase pathway which produces immunoneutral compounds that promote tissue healing. Diets with an excess of 6 omega to 3 omega fats have resulted in documented

population increases of allergies, autoimmune thyroiditis, juvenile rheumatoid arthritis, inflammatory bowel diseases (Crohn's and ulcerative colitis), asthma, atherosclerotic heart disease, and systemic lupus.

Before we get carried away and become strict vegetarians and clean out the health food stores of all the 3 omega fatty acid bottles, it is important to emphasize the word balance. It is not that 3 omega fats are good and in contrast 6 omega fats bad; they are both important and essential to our health. A diet deficient in 6 omega fats would result in easier injury, poor recovery from injury, and decreased efficiency of our immune system against infectious disease. In fact, several athletes who became strict vegetarians and threw their diets out of whack in the 3 omega direction, noted an increase in severe injuries, most notably ligament tears and fractures. The 6 omega fats are vital for tissue strength and bone integrity. Both 6 omega and 3 omega fats are essential (meaning required) in our diet. Perhaps the Bible says it best: "All things in moderation."

The major sources of 3 omega fats in the American diet are from fish and vegetable oils. EPA and DHA are the 3 omega fats that are being added to baby formulas. They are predominantly available in our diet by eating fish. Vegetables are high in the 3 omega, ALA, but although humans can synthesize EPA and DHA from ALA we are not very efficient at it. In addition, if we are consuming a lot of 6 omega fats we inhibit the synthesis of DHA and EPA. So it behooves us to eat adequate DHA and EPA. The preceding chart from Kris-Etherton et al. gives the percentage of 3 omega fatty acids by weight from fish commonly available in the US.

Seafood	n-3 Fatty Acids
	% by weight
Mackerel	1.8-5.3
Herring	1.2-3.1
Salmon	1.0-1.4
Tuna	0.5-1.6
Trout	0.5-1.6
Halibut	0.4-0.9
Shrimp	0.2-0.5
Cod	0.2-0.3
Plaice, Flounder and Haddock	~0.2

Vegetable oils that are high in the 3 omega fat, ALA, are flaxseed, soybean and canola oils. By changing the way we cook (using the above oils for frying), by adding green leafy vegetables, fruits and nuts to our diet, and by increasing our consumption of fish and leaner meats, we can bring our diet back into balance, be healthier, recover faster, and perform up to our capabilities.

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disorder when you see a parent whom you experience as hostile, dependent, complaining, demanding, or helpless in the office, ward, or emergency department. Attempts to help often lead to increasing frustration, and the staff often feel overburdened by this parent. The term 'borderline', however, may be applied indiscriminately to parents who cause the staff difficulty. By properly identifying these parents, the hostile feelings generated within the medical staff may give way to a more empathic viewpoint, and lead to more effective management tools.

Children in these families often have psychological difficulties, and up to 24 percent have witnessed a suicide attempt in their parent. As two percent of the general population has BPD, pediatricians have little hope of avoiding them. In this article we hope to give the medical staff the ability to recognize and manage parents with borderline personality disorder.

Etiology

A biopsychosocial model proposed by Paris suggests a complex interaction of factors in the development of BPD. Poorly understood biological factors, especially difficult temperament, are a required substrate. Additional psychological factors are sexual trauma (often early, prolonged, severe, incestuous), early separation or loss, and emotionally unattuned or deficient parenting (parents with alcoholism, depression, or personality disorders). Finally, the normally protective family social structure is disrupted. Since a series of interactions appears to be necessary to develop BPD, interrupting the chain may either prevent or ameliorate this disorder. Conversely, societal changes causing family breakdown and decreasing parental availability may lead to increased incidence of BPD.

Diagnosis

Initially these patients were thought to be borderline schizophrenic, because they may briefly become psychotic (lasting a few hours) in certain circumstances, such as facing perceived abandonment. Later they were thought to be on a border between psychosis and neurosis.

More recently the DSM-IV considers BPD to be one of the personality disorders. The DSM-IV criteria for BPD, ranked in order of importance, requires five of the following nine criteria:

1. Frantic efforts to avoid real or imagined abandonment.
2. A pattern of unstable and intense interpersonal relationships characterized by alternating between extremes of over-idealization and devaluation.
3. Identity disturbance: markedly and persistently unstable self-image or sense of self.
4. Impulsivity in at least two areas that are potentially self-damaging, e.g., spending, sex, substance abuse, shop lifting, reckless driving, binge eating.
5. Recurrent suicidal threats, gestures, or behavior, or self-mutilating behavior.
6. Affective instability due to a marked reactivity of mood (e.g., intense episodic dysphoria, irritability, or anxiety usually lasting a few hours or rarely more than a few days).
7. Chronic feelings of emptiness or boredom.
8. Inappropriate, intense anger or difficulty controlling anger, e.g., frequent displays of temper, constant anger, recurrent physical fights.
9. Transient, stress-related paranoid ideation or severe dissociative symptoms.

Accurate diagnosis of BPD may be difficult because of imprecision in the diagnosis, and the large differential diagnosis of other personality and psychiatric disorders. However, identification of key BPD symptoms may be sufficient for the pediatrician to develop a plan of appropriate interactions. After interviewing a parent, some clues that should lead you to consider BPD include:

- An inability to form a coherent picture of the parent, their situation or the people in their lives;
- Instability in relationships, work, school;
- You are praised inappropriately, while others are condemned;
- Problems are not verbalized well;
- You often feel emotionally drained and empty, which may serve as a clue to the patient's unspoken or diffusely presented subjective experience of the world.

Practical Difficulties

The kinds of difficulties the medical staff is likely to experience from the borderline parent include:

1. Intense, hostile, negative affect on the parent's part that strikes a strong, negative counter-response from the medical staff.

2. Frequent use of primitive defense mechanisms which are usually seen in infants or young children. Toddlers experience a barrage of feelings that they can't differentiate significantly, and have difficulty organizing their feeling states. These manifest in the adult as:

- Affective storms of rage, or adult versions of tantrums, to deal with their own fragmented identities.
- Splitting others into all good (overvalued) or all bad (devalued) personalities. This results in pitting

the 'good' staff against the bad staff, and even switching back and forth. Statements or other indications from the parent that you are the best doctor (or nurse) who has taken care of their child are often encountered. In the most troubling instances the staff actively participates in this split without realizing it has occurred.

- manipulation.
- These evoke in the doctor feelings of both helplessness and punitiveness which the parent experiences but cannot handle.

3. Extreme fear of separation or abandonment, to the point of viciously attacking you, if the parent believes you might no longer want to see them. This may occur if you go on vacation, or fail to tell them when to return for their next visit. Distortions of truth, including psychotic ideation, are often experienced by the parent when abandonment is perceived. They strike out in order to force you back to them. They try to win you by complaining about you.

4. Chronic emptiness, boredom, or dysphoria.

5. Impulsivity, including sexual indiscretions. Seductive behavior and dress are not uncommon in borderline individuals, and may represent their attempt to demonstrate how they value you, as well as to ensure your attachment to them. The unwitting provider may buy into this, brought long by their desire to rescue the parent from their awful situation. During or after a brief psychotic episode, they may falsely accuse you of sexual harassment to abort a perceived abandonment.

6. Aggression, either self-directed (mutilation or suicide) or against others (ranging from verbal abuse to homicide). The completed suicide rate is 10%, and other early accidental death is 8% more.

7. Poor social adaptation, with limited ability to follow usual social customs.

8. Extreme and inappropriate demands, such as seeing you without an appointment ahead of other patients, or standing in your doorway until you see them.

9. Lying to achieve a particular goal (e.g., about adverse life circumstances, illness, abuse, etc.)

Management of Borderline Personality

Knowing about BPD is the first step, and leads to a number of ways to help your staff and the families of these parents.

1. Since separation is a primary threat, provide direction and structure. These are also termed boundaries

a. Tell them when you want to see them again. A scheduled appointment within a reasonable time will lessen the in-between disruptions.

b. Give them firm guidelines regarding how each of them is expected to conduct himself/herself. For example, you might need to set limits on walk-in appointments, or how many problems per visit are allowed, or how long you are allowed to spend with them.

c. A written contract may be helpful to both provide structure and limits.

d. Clearly establish who is the child's care provider (with perhaps one alternate), as well as who the child's nurse is (if admitted).

2. Educate your staff about borderline parents. Head off splitting by meeting with your staff as often as needed, or anyone else involved in the child's care, to discuss problems. This will diffuse erroneous information, as well as inform everyone about the rules of conduct for the parent.

3. Develop a team approach with other staff. New ideas may be generated. A consult to the psychiatric consultation service is often extremely helpful to head off problems among the staff.

4. Avoid the accusation of sexual misconduct. Sometimes these parents are sexually inappropriate, at other times their psychotic thinking may lead them to make false accusations regarding your behavior. Occasionally physicians and other providers have been guilty of having sexual relations with patients. Furthermore, these patients may lie to hurt you or prevent you from leaving them. Having a witness present during the exam, and keeping the door open during the evaluation, may prevent later difficulties in this area. Giving in on this issue is often career-ending. Clearing your innocent name may be difficult or even impossible.

5. Document patient encounters appropriately, and protect the medical records by keeping a separate copy. Tape recording the visits by a borderline parent, with their written consent, may be required to protect you.

6. Affirm the parent that both you and they have the child's interest at heart, and that they have a right to the best care available. Avoid attacking their sense of entitlement. When possible, point out what the parent is doing positively with the child. This may then be linked to your plan for the child. For example, you might say: "Ms. J., your child is entitled to the best medical care available. However, since your child does not need a brain MRI, obtaining one would be bad care for your child." Another example could be: "Mr. Y., you have

been doing a great job following your child's complex medical problems. The next thing I want you to do is to call before coming in so that I can have enough time to spend with you."

7. Immediately report suicidal threats to appropriate medical services, as almost ten percent of borderline individuals do kill themselves.

8. Think about child abuse or neglect, and don't delay reporting it, although this will cause your relationship to acutely deteriorate. Some BPD parents, especially those with a medical background, may harm their child by making him ill, or fabricating an illness (Munchausen's by proxy).

9. Recognize your own negative feelings toward the parent (counter transference), as well as your own rescue fantasies for the child.

10. Refer these parents for psychological help. Long-term psychotherapy and medications (antidepressants, mood stabilizers, and neuroleptics) have been demonstrated to significantly help these individuals, and should help the families and yourself. Although the parent might not go, you will have done what is best for the family. The best news is that the majority of these parents over many years will no longer meet the criteria for borderline personality disorder. Hanging in there is good for the children and the family.

Summary

Many of these parents do improve in their ability to interact with others, though the process may take years. They will not disappear from your practice. If you learn ways to more appropriately manage them, life will be more enjoyable for everyone involved.

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Small for Gestational Age Infants: Risks Beyond the NICU

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Introduction

Most of us are aware of the immediate health concerns of the small for gestational age (SGA) newborn. These babies may have problems including low Apgar scores, meconium aspiration, hypoglycemia, pronounced physiologic hypocalcemia of the newborn, respiratory difficulties, hypothermia, feeding difficulties and an increased incidence of future failure to thrive. Less commonly known are the long-term risks of SGA babies. SGA has been associated with several risk factors later in life. Among these are hypertension, obesity, glucose intolerance, type 2 diabetes, dyslipidemia, polycystic ovarian syndrome, premature adrenarche, precocious puberty, decreased male and female fertility and adult short stature. While the pathogenesis of each of these conditions has not been fully explained, alterations in insulin and follicle stimulating hormone (FSH) sensitivity appear to be the most likely pathogenesis. As these conditions often do not express themselves until adolescent or adult life, the link to former SGA has not been identified until recently. Additionally, for those SGA babies who do not exhibit catch up growth by 2 years of age, final adult height often is less than mid parental height would predict. Recently, growth hormone has been FDA approved for treating this group of SGA patients. Early identification of SGA infants at risk and appropriate referral is important for optimal outcome of these patients.

Insulin Resistance

Epidemiological studies have shown an association between SGA and future development of type 2 diabetes. This association has been described in U.S., U.K , European and Taiwanese studies. The mechanism for this association has not been identified. However, several studies have suggested that the underlying pathogenesis may be impaired insulin secretion and insulin resistance. Recently, Veening, et al[1] studied a cohort of 28 Caucasian prepubertal SGA children and 22 age matched AGA controls.

SGA babies are at risk for development of obesity, insulin resistance, PCOS, precocious puberty, hypertension and

Insulin sensitivity was measured using the hyperinsulinemic-euglycemic clamp technique. She found that insulin sensitivity was significantly lower in SGA children compared with controls. However, insulin secretion was found to be normal. Autopsy studies by Beringue, et al[2] in 21 SGA infants matched to a cohort of 15 infants whose weight was between the 50th and 75th percentile over an eight year period failed to show a relationship between islet cell density and fetal weight. Obviously, functional studies could not be done, but other studies suggest that insulin secretion in SGA neonates is normal. Bazaes et al[3] studied 136 SGA breastfed infants and 34 AGA controls at 48 hours of age and was unable to demonstrate a difference in insulin sensitivity. He proposed that "adiposity rebound", as SGA babies are at increased risk for future obesity, may account for the later development of insulin resistance in these infants. Ongoing studies in the U.K.[4,5] are prospectively studying the combined effects of SGA and weight gain on sensitivity to insulin.

Hypersecretion of FSH

SGA has been associated with hypersecretion of FSH and reduced gonadal growth in adolescent boys and girls. SGA effects on premature adrenarche and insulin sensitivity have been evident before the onset of puberty. Ibanez et al[6] assessed serum concentrations of FSH and inhibin B in 46 infants, 17 AGA and 29 SGA, at a mean of 4 months of age, before the physiological decrease in activity in the gonadal axis. In SGA girls and boys, FSH levels were 2 and 4 fold greater, respectively, than in AGA controls. Inhibin B levels were similar in both groups. SGA infants appear to require an augmented FSH drive in both sexes to maintain the reproductive axis in both sexes. It remains to be studied as to whether FSH hypersecretion in infancy is a marker of subsequent subfertility. In the female fetus, IUGR has been associated with a reduced fraction of primordial follicles[7]. A unifying hypothesis would be that poor fetal growth results in reduced amounts of granulose and Sertoli cells within the developing gonads, leading to a decreased secretion of Inhibin B on the feedback side of the hypothalamic pituitary gonadal loop. This would require an augmented FSH secretion to drive the efferent side of the loop. The role of FSH hypersecretion in the pathogenesis of polycystic ovarian syndrome has not been resolved. However, an increased prevalence of polycystic ovarian syndrome in former SGA infants may lie in this enticing early disturbance of the hypothalamic pituitary gonadal axis.

SGA babies may be at future risk for subfertility.

Growth Hormone

Most SGA infants experience catch-up growth and achieve a height above the 2.5 percentile on the growth chart by 2 years of age. A small fraction may continue catch-up growth and not attain the 2.5 percentile until 3 years of age. For those who have not regained normal height by 2 years of age, growth hormone administration has been approved by the FDA in July 2001.

The International Small for Gestational Advisory Board[8] consensus statement on the management of short children born small for gestational age recommended referral of those children who did not catch up by 2 to 3 years of age to a pediatric endocrinologist for assessment. The goal was to achieve catch up growth in early childhood, maintenance of normal growth in childhood, and achieve a normal adult height. Pomerance[9] raises appropriate concerns as to the wisdom of universally accepting these recommendations. He sites the lack of data demonstrating an improved quality of life or improvement in self-esteem in children treated with growth hormone for idiopathic short stature. Outcome data is limited and conflicting as to the attainment of final height. Finally, he questions whether third party insurance will reimburse for expenses incurred as there is no information on projected savings to society. A red flag is raised by Silverstein and Shulman[10] on an increased incidence of type 2 diabetes in SGA growth hormone treated patients. The already known association of SGA and increased type 2 diabetes risks makes the routine treatment with growth hormone worrisome. The military is currently working on a policy to address the treatment of idiopathic short stature. SGA indications will likely be included.

Summary

While the immediate risks to the SGA newborn are well-recognized, anticipated and managed by the general pediatrician, long-term risks of SGA birth are less well-recognized. Growth and development of these infants needs to be continuously assessed. Early anticipatory guidance of parents of these infants is important. Early recognition and referral for poor catch up growth by 2 years of age is warranted. While

SGA babies may not catch-up by 2 years of age; endocrine referral for assessment for growth hormone treatment is warranted.

not without risks, toddlers treated early with growth hormone exhibit better catch up growth than older children and adolescents. Careful monitoring for side effects of hyperglycemia and obesity needs to be undertaken. Caretakers need to be aware of the risks of early weight gain on future obesity, type 2 diabetes, dyslipidemia and hypertension. The SGA infant may be an especially at risk category in the current "obesity epidemic". The association of insulin resistance, obesity and SGA in future polycystic ovarian syndrome may also be amenable to early identification and counseling of those at risk.

Ongoing research will more clearly identify the pathogenesis of these associations. Early identification of factors that predispose to SGA infants, such as cigarette smoking, may be another place where we, as pediatricians, can help parents prevent future occurrences of SGA.

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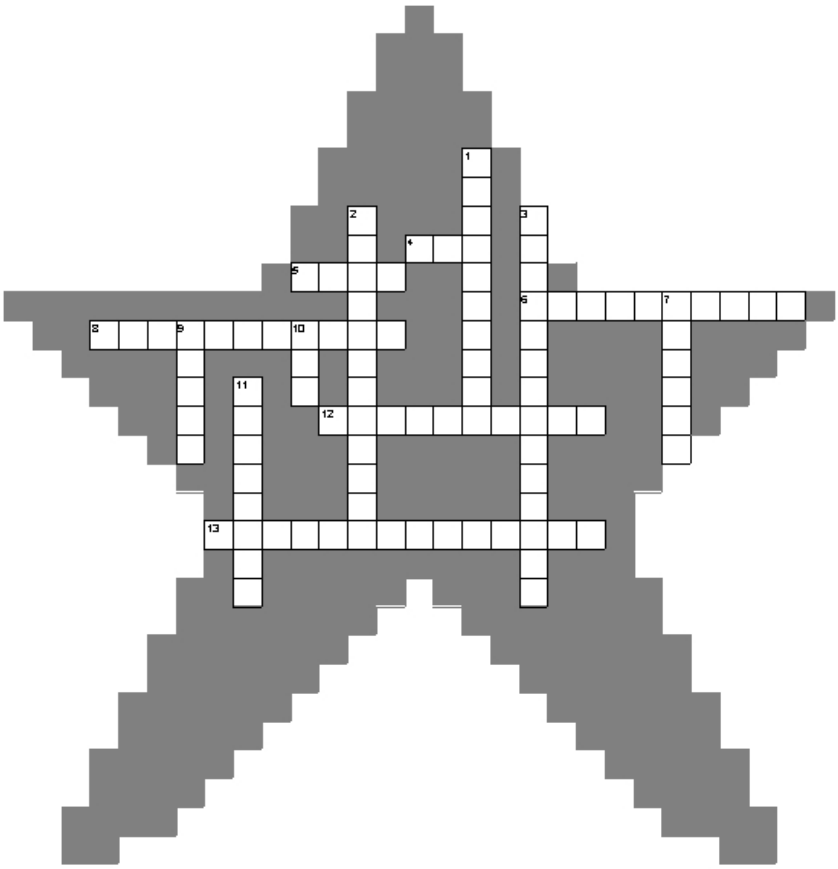

Word List

adrenarche
amenorrhea
canola
cytoplasmic
depolarization
dyslipidemia
euglycemic
fish
hypersecretion
omega
SGA
ten
Wegeners

Across

4. percentage of borderline personality disorder patients who are successful in their suicide attempt
5. predominant source in our diet for 3 omega fatty acids
6. the state of normal blood glucose concentration
8. antineutrophil _____ antibodies directed against proteinase 3 are specific for Wegener's Granulomatosis
12. the physiologic change at puberty caused by adrenocortical secretion of androgenic hormones
13. method by which a nerve signal travels down a nerve

Down

1. a significant side effect of cyclophosphamide therapy in female patients
2. a disorder of lipid metabolism
3. SGA has been associated with this effect on FSH and reduced gonadal growth in adolescent boys and girls
7. a vegetable oil in addition to flaxseed oil and soybean oil that is high in 3 omega fatty acids
9. type of fatty acid essential for the formation of every cell in our body
10. long term risks for these size affected infants have included obesity, hypertension, glucose intolerance, Type 2 diabetes, dyslipidemia, polycystic ov
11. a granulomatous disease marked by systemic vasculitis of both medium and small arteries