

PEDIATRIC NEWS

San Antonio Military Pediatric Center



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Housestaff Puzzler

Benign Rolandic Seizures

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A 6 year old Hispanic male presented to the neurology clinic for follow up for his seizures. He had been treated with Tegretal for two years for his seizures and had not had a seizure since starting medication. As he was a new patient to the service, his initial history was elicited. Per his mother, at 4 years old, he started having nocturnal seizures consisting of grunting noises and shaking. The first seizure was noted when he was sleeping in his parents bedroom. It lasted approximately ten minutes after his parents woke and found him shaking all extremities. His eyes were closed at the time. It is unknown whether he had enuresis. He was taken to the emergency room, where he was fully arousable but sleepy. The evaluation included a lumbar puncture and CBC, both of which were normal. After his

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The Prenatal Visit: Chore or Opportunity

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The prenatal visit is one of the health supervision recommendations from the Academy of Pediatrics least emphasized during Pediatric residencies, yet it may be one of the most useful visits in establishing a relationship between a family and their pediatrician. In private practice, a prenatal visit may range in scope from a five minute phone call discussing office hours to an hour group visit involving several families. Ideally, the prenatal visit will allow an opportunity for a new family to get to know their pediatrician well before the excitement and stress surrounding the birth of their baby.

Given the demands of a busy general practice, why should a pediatrician even consider taking the time to meet with a prospective family? The prenatal visit should serve as an opportunity to establish a physician-parent relationship, gather history about the family and provide information ranging from how the office is run to advice about care of the newborn infant. In addition, a prenatal visit can help the new parents to develop a sense of competency. The majority of referrals of new patients in private practice come from obstetricians and a well thought out prenatal visit is an effective marketing tool. The arrival of a baby is one of the most important events in parents' lives. Caregivers who have the opportunity to meet face to face with a pediatrician and receive answers to questions will leave with a sense that they

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Opioid Tolerance and Dependence in Critically Ill Infants and Children

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Introduction

Providing adequate sedation and analgesia in the intensive care setting is driven not only by humanitarian goals, but also by the accumulation of evidence that details the biochemical and physiological stress response in infants and children to critical

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first seizure, he had several more nighttime seizures during sleep, once every month or so, lasting up to 15 minutes at a time. After pediatric neurologic evaluation, he was placed on low dose Tegretal (carbamazepine), which suppressed his seizures completely. His past medical history was benign, with no hospitalizations or surgeries. Developmentally, he had walked at 11 months, had distinct words at 13 months, and was doing well in first grade. The patient's mother was not concerned about his intellectual functioning as compared to his peers. Family history was significant for an uncle who also had seizures as a child, but none as an adult.

The seizure disorder that this child's history suggests is benign rolandic epilepsy (BRE). BRE is solely a childhood epilepsy syndrome. Seizures mostly occur at night and usually have a brief focal onset that can evolve into generalized tonic clonic seizures. Children can also have daytime seizures that are partial seizures, often involving one side of the body, that usually do not become generalized. The face is frequently involved, possibly the same side arm and leg as well. If the child is awake, they are often aware of the seizure and may walk into their parents' room in the middle of the seizure. BRE occurs between the ages of 2 and 16 years old, most commonly between 5 and 10 years old. It does not persist past the second decade. Children usually have normal development and normal neurologic exams. Nocturnal seizures rarely wake children from sleep and daytime seizures rarely impair consciousness. There is infrequent post-ictal confusion or amnesia. Seizures in BRE are distinct from other types of partial seizure disorders, such as temporal lobe epilepsy, as there are no

automatisms, auras, illusions, hallucinations or affective symptoms. BRE has a benign prognosis, as evidenced by the title. There is no increased risk of lifetime epilepsy with BRE and no resultant neurologic abnormalities. However, some studies have shown that there is an increased incidence of learning disorders in children with a history of BRE.

This patient's presentation was relatively typical. Parents usually do not recognize seizures unless the child is sleeping in the same room. Often, this occurs during vacations or family trips. The focal presentation is usually missed, but the generalized tonic-clonic movements and guttural vocalizations wake the parent from sleep.

There is often a family history of BRE, although the parents may not be aware of seizures that occurred only during childhood. Inheritance is autosomal dominant with age-dependent penetrance. Fifty percent of first degree relatives have abnormal EEG finding between the ages of 5 and 15 years old. However, only 12% of children with these EEG findings have clinical seizures symptomatic of BRE.

Diagnosis of BRE is based primarily on clinical history and EEG findings. The clinical history has been discussed in the above text. EEG findings consist of a normal background with superimposed high-amplitude spikes, followed by a prominent slow wave. The spike-wave complexes occur unilaterally or bilaterally over the midtemporal and central (rolandic) regions of the EEG. EEG findings can be found awake, during sleep, or both. Progression of the EEG to focal slowing is not characteristic and should alert the clinician to other causes of epilepsy.

If a patient that you suspect has BRE has diagnostic EEG findings and clinical history, coupled with a normal neurologic exam, no further work up is necessary. If the EEG is not diagnostic, the clinical history is

unclear or there is an abnormal neurologic exam, further evaluation is necessary. An MRI should be performed to rule out a focal lesion as a source of partial seizures.

Treatment for BRE is initiated only after several seizures have occurred, as many children may only have one or two recognized seizures in their lives. Further, these seizures have minimal associated morbidity, as seizures usually occur during sleep and status epilepticus is exceptionally rare. If treatment is needed, a single anti-epileptic drug (AED) usually suffices. Medicines used to treat partial simple and partial complex seizures are effective. If medications are used, nearly any AED will be effective, but carbamazepine is the most commonly used. Several studies promote the use of Neurontin monotherapy due to the low side effect profile. Oxcarbazepine is also an attractive alternative for similar reasons. Many parents are comfortable foregoing treatment with medications unless seizures are frequent, severe or diurnal. If AED therapy is used, treatment for 1-2 years is recommended before tapering. It is reassuring if the child has a normal EEG prior to tapering, but poorly correlated to seizure recurrence. A child with a normal EEG has a slightly lower risk of recurrence, but seizures might still recur. Therefore, EEG is not necessarily performed prior to discontinuing medications. If one medication is unable to suppress seizures, it is prudent to perform further evaluation including CT or MRI to evaluate for other etiologies.

To summarize, BRE is one of the few seizure disorders that, although frightening to parents, has few complications. Seizures generally occur at night, limiting injury to the child, and it is extremely rare for status to occur. Evaluation should include a neurologic exam and EEG. With classical findings and few seizures, many children can be

managed without anti-epileptic drugs, limiting side effects. For those children who need medication, it is usually for 1-2 years and seizures regress of their own accord. As general pediatricians, it is helpful to feel comfortable with BRE in order to offer support and information to parents of these children.

Resources

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will be able to relate to that pediatrician. A hurried superficial visit will send them elsewhere.

Parents who benefit from prenatal visits are first time parents, families who are new to an area or a practice, parents experiencing a high risk pregnancy, families about to adopt and families who may have experienced a perinatal death. A prenatal visit may be organized in a variety of ways but should contain some basic exchanges of information both from the physician and from the expectant parents. At the outset of the visit, parents can be given a registration form or health questionnaire that can be read prior to the physician talking with them. Brochures with information about the practice and patient education handouts can be given to the parents to read while waiting for the physician to begin the interview. Some practices show educational videos about breastfeeding, circumcision or safety prior to the interview. At the end of the interview, a summary of the information ex-

changed and an opportunity for questions helps tie up loose ends.

Many practices have printed brochures to give to parents that cover office hours, how to call after hours, which hospitals are affiliated with the practice and important phone numbers. Some brochures include pictures of the providers and a summary of their credentials. Parents need to know who will answer their calls both during and after office hours and the time frame for returning calls, as well as how to contact the practice in case of an emergency. Parents will want to know how soon after birth their baby will be examined and which provider or providers cover the practice when their pediatrician is not available. Parents like to have information about the fee schedules of the practice, although now most of the information about copays for office visits, medications and procedures comes from the insurance carrier. Physicians can suggest to families that they contact their insurer with those questions, as many patients do not know what their portion of the expenses will be.

A physician should ask about the pregnancy. The mother's feelings about the pregnancy, any medical problems that are pertinent and a history of medications should be elicited. Questions about congenital illnesses or hereditary conditions, hypertension, diabetes, heart disease, asthma or sickle cell disease can be asked at this time. Direct questions about tobacco and alcohol use as well as any history of substance abuse can be included as part of a family medical history. If positive, this is an opportunity for anticipatory guidance and referral of parents for help with substance abuse issues. Some practices use this opportunity to review the mother's Hepatitis B and Group B Streptococcal status as well as the RPR and HIV status and keep the

information on file for nursery rounds and follow up. If there are known complications with the pregnancy, this is an opportunity to discuss them and help parents know what to expect after the birth in the way of testing or interventions. This can also be a time for the pediatrician to share his or her experience in dealing with these types of problems.

A social history is an important part of the prenatal visit. Questions to consider are the parents' childhood experiences, exposure to abuse, educational levels and expectations about the child and parenting. Discussions about resumption of work or school and child care options can be helpful. Support systems both within the home and outside the family can be discussed including a reminder to plan for respite for the primary caretaker. As more people from other cultures come into the United States, a sensitive review of any traditional medical practices or beliefs may be helpful in preventing problems with communication in the future. In many cultures, grandparents or family elders play a significant part in raising the baby, their roles should be explored if applicable.

The labor and delivery should be discussed. Prenatal classes can be recommended if not already attended. A history of previous labors including any complication or postpartum depression should be elicited. The importance of the father's participation in labor and delivery can be discussed. If a father is unavailable, the mother should be encouraged to have a support person in attendance at the birth. Discuss the option of rooming in if available and resources available to the family after discharge. Reviewing the time frame for newborn exams, labs or immunizations to be done in the nursery can be helpful as well as follow up to any

procedures or labs that may be necessary.

Nutrition is an important topic that must be discussed at the prenatal visit. For families who have not yet decided, a discussion of breastfeeding and its' advantages can take place. Families who have elected to bottle feed should know which formulas are available and how to prepare them. For mothers who decide to nurse, resources such as a lactation consultant or other support groups can be made. Any family history of milk intolerance or allergy should be discussed. Many parents change formulas or stop breast feeding in the first few weeks based on perceived symptoms of milk allergy or intolerance. A brief review of normal feeding and stooling patterns may be helpful at this time. If nothing else, the suggestion to discuss feeding changes or problems with the physician prior to implementing changes may avoid discontinuation of breast feeding or multiple formula changes.

If a male infant is expected a decision about circumcision should be made prenatally. The potential risks and benefits can be discussed as well as the use of local anesthesia. The type of procedure and follow up care necessary can be reviewed. In some hospitals the obstetrician performs the circumcision, in others the pediatrician provides this service, parents should know what to expect. Reminding families that circumcision is an elective procedure can be helpful especially if there are complications in the nursery that would preclude the procedure such as sepsis, hypospadias or other medical conditions.

Sleeping arrangements and positioning of the baby in the supine position for sleep should be

reviewed. Co-sleeping may be a cultural or desired practice for the family, reminders about soft sleeping surfaces or the risk of rolling over an infant during heavy sleep should be discussed. The discussion of sleeping face up in order to decrease the risk of SIDS should also include changing the sleeping position to avoid positional plagiocephaly, "tummy time" while awake and avoidance of blankets and stuffed toys. A review of crib safety can be done at this time.

Other safety issues that should be discussed include car safety and car seats, home safety and CPR training. Remind parents to have the car seat available for the first ride home from the hospital as many hospitals will not discharge infants home without one. The review of smoke detector placement in the home and the temperature of hot water heaters will begin to help parents think of safety issues that they may not have considered.

Parents may have questions about child rearing. If there are older siblings in the home a discussion about sibling rivalry and possible regressive behaviors can be helpful at this visit. Pointing parents to written resources and reminding them to include the older sibling in preparing the home for the new baby is helpful. Reassuring parents that regression to infantile behaviors is expected and normal will aid in the transition time for the family.

A prenatal visit can be structured in a variety of ways and cover a broad spectrum of information. While the structure and information will vary from practice to practice, the fundamental concept of the pediatrician as a partner to the parents in raising a healthy child in a safe environment is a consistent one. Most of us enter pediatrics with this concept in mind. A prenatal

visit can help parents to see that about us and encourage them to use us as allies in their adventure of parenting.

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illnesses and major surgical trauma. These stresses and the patient's response to the stress may directly influence the patient's outcome. The modification of the stress response remains the central component of modern day anesthesia, the administration of systemic opioids and regional anesthesia are the favored routes for modification.

The benefits of stress-reducing opioid therapy have been demonstrated in the critical care setting many times. Anesthesia or continuous analgesia with fentanyl have both been associated with improved survival in neonates with congenital diaphragmatic hernia and with decreased pulmonary vascular responsiveness in infants following repair of congenital heart lesions. Following the repair of congenital

heart lesions, neonates who had high-dose opioid anesthesia followed by postoperative continuous opioid infusions, have decreased concentrations of stress hormones and to have improved clinical outcomes (i.e. less postoperative infections, DIC, persistent metabolic acidosis and more importantly, increased rates of survival to ICU discharge).

The prolonged or frequent use of opioid in the critical care setting is frequently associated with the development of tolerance and dependence to these drugs, leading to the narcotic abstinence syndrome (i.e. *narcotic withdrawal*) when the opioids are stopped. Opioid withdrawal can be prevented with appropriate weaning schedules, diagnosis/identification of withdrawal by objective clinical methods, and the treatment by a variety of pharmacologic and non-pharmacologic means.

Misconceptions

1. Neonates lack the neurologic substrate for the perception of pain.
2. Even if neonates experience pain, they have no memory of it.
3. Even if neonates experience pain and remember it, it is not detrimental to them.
4. It is too dangerous to administer anesthesia or postoperative analgesia to neonates.

Definitions

Pain—An unpleasant sensory and emotional experience associated with actual or potential tissue damage. (International Association for the study of Pain)

Agitation—Excessive gross motor activity and crying

Tolerance—Neurochemical adaptation of neuronal cells (not a change in metabolism) where

following frequent drug dosing, there is a decrease in the pharmacological effects, an increased dose is required to produce the same physiologic effect.

Cross tolerance—Phenomena where the tolerance to one agent causes in some degree of tolerance to the exposure to a different agent.

Physical dependence—Altered physiologic state resulting from the chronic administration of a drug. Unless there is continued administration of the drug, withdrawal symptoms may occur. Can occur with or without addiction.

Addiction—A complex behavioral pattern of drug use, characterized by an overwhelming involvement with the compulsive use of the drug, the securing of its supply, and a high tendency to relapse after withdrawal. Physical dependence is not necessarily a feature of addiction.

Withdrawal/Abstinence Syndrome—A constellation of symptoms chronologically following the abstinence or decreased utilization of the drug the person has become dependent (physically) upon. The symptoms were not present prior to the initiation of the drug, should evolve and resolve over a well-defined period of time, and can resolve upon the reinstatement of the appropriate amount of the drug.

Clinical Features

Neonatal Abstinence Syndrome—Term utilized to describe a constellation of symptoms exhibited by infants born to mothers who are opioid dependent during pregnancy, first described by Finnegan in 1975. Little literature exists addressing the problem of therapeutically induced (iatrogenic) opioid withdrawal.

Neurologic excitability—jitteriness or tremors, hyperactive reflexes, hypertonicity, irritability, decreased sleep or frequent awakenings, high-pitched and excessive crying, frantic nonproductive sucking and rooting behavior.

GI dysfunction—poor feeding, vomiting, and diarrhea.

Respiratory distress—Tachypnea and sneezing.

Autonomic hyperfunction—Sweating, nasal stuffiness, fever, and mottling.

Studies in both animals and humans have shown the rapid development of tolerance when narcotics are administered by continuous infusions. It was previously assumed that withdrawal usually occurs only after prolonged exposure to opiates.

Tolerance to fentanyl rapidly occurs when used as a continuous infusion. Fentanyl plasma levels of 7.7 to 13.6 ng/ml (mean 10.6 ng/ml) are considered to provide satisfactory anesthesia for PDA ligation. After 6 days of continuous infusion (in patients on ECMO), mean fentanyl plasma concentrations of 13.9 ng/ml were needed to achieve a moderate level of sedation, with augmentation with benzodiazepines.

Fentanyl is widely reported to be approximately 80-100 times more potent than morphine as an analgesic, based on single dose administration. But, anecdotal evidence suggests that fentanyl has less sedative effect than morphine does.

Patients sedated with fentanyl are almost twice as likely than those sedated with morphine to develop narcotic abstinence syndrome after discontinuation of therapy.

Total fentanyl dose and length

of infusion have been shown to be significantly greater in infants who experienced withdrawal. The peak infusion rate has not been shown to be significantly different.

Fentanyl dose of 1.5mg/kg or duration of infusion of 5 days is associated with a >50% chance of developing withdrawal.

Fentanyl dose of >2.5 mg/kg or a duration of infusion of >9 days is 100% predictive of withdrawal.

Benzodiazepine withdrawal differs marginally from opioid withdrawal. Benzodiazepine withdrawal is usually not associated with gastrointestinal features. Classical features are severe anxiety, tremors, confusion, insomnia, perceptual disorders, depression, and generalized seizures. Movement disorders may persist for 6-8 weeks after discontinuation of midazolam and are poorly responsive to treatment with other benzodiazepines.

Treatment

Supportive nursing care to reduce irritability, to promote feeding and sleep, and prevent or minimize complications from hyperactivity or diarrhea is the mainstay of therapy for mild symptoms. Supportive care,

monitoring, and treatment of other medical conditions are important.

For Neonatal Abstinence Score of <8 comfort measures and decreased environmental stimuli are recommended.

Pharmacological therapy is usually initiated for NAS ≥8 for 3 consecutive scores (2 hours between assessments) or for 2 serial scores >12.

Single agent therapy should be used whenever possible.

Conversion

(Fentanyl mcg/kg/hr X wt.)60 = IV dose MSO4 q4hr or Methadone dose

IV dose of methadone X 2 = oral methadone dose (75% bioavailability)

Methadone dose q12 hr X 3 doses (loading) then wean 10% q day for 10 days

Morphine

Oral dose of 2-6 times the IV dose can be used to treat withdrawal

Methadone

Long duration of action, IV or PO routes.

Clonidine

a-2 adrenergic agonist that also activates the same K-channel as the m-opioid, so it may “mimic” the opioids at the subreceptor level.

Shown in adults and animal studies to suppress many of the autonomic symptoms of opioid withdrawal.

NO effect on GI symptoms, less effective than opioids for treatment of sleep disturbances. May produce hypotension and sedation.

Dose: 3-5 mcg/kg/dose. Dosed q6 hr.

Weaned over 3 days once off of methadone (when used together).



**What Is That Hole For???:
What the pediatrician should know about urinary reconstruction**

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As the number of children with some form of urinary reconstruction continues to grow, it is likely that you will encounter these patients in your clinical practice. Since the elements of reconstruction can vary from patient to patient, some confusion about exactly what lies beneath the stomas and scars in your patient is natural. The purpose of this article is to provide a basic understanding of urinary tract reconstruction and potential long-term complications to

Neonatal Abstinence Score

Sign/Symptom	Score	Sign/Symptom	Score
Cry		Sweating	1
Excessive	2	Fever	
Continuous	3	100-101	1
Sleep		>101	2
<1 hour	3	Mottling	1
<2 hour	2	Nasal stuffiness	1
<3 hour	1	Sneezing	1
Moro reflex		Nasal flaring	2
Hyperactive	2	Respiratory rate	
Markedly hyper	3	>60	1
Tremors		>60 & retractions	2
Mild disturbed	1	Excessive sucking	1
Mod-severe dis	2	Poor feeding	2
Mild at rest	3	Regurgitation	2
Mod-severe at rest	4	Vomiting	3
Hypertonicity	2	Stools	
Frequent yawning	1	Loose	2
Excoriations	1	Watery	3
Seizures	5		

help you identify problems and encourage compliance in your patients.

Long-term objectives in the medical and surgical management of children with neuropathic bladder and other congenital and acquired anomalies of the urinary tract include: 1) achieving social continence while 2) preserving upper tract function. Historically continence was not an option and many of these patients either remained incontinent, wore a catheter long-term or had the urinary stream permanently diverted. An *ileal conduit* to permanently divert the urine to an appliance on the abdominal wall remains a common option in the adult patient who requires removal of the bladder, but is now performed in children only in truly exceptional cases. The ileal conduit is clearly undesirable for most young patients, as it unnecessarily carries the stigma of an incontinent stoma and does not uniformly result in preserved renal function over many years. This is also not a good temporary measure since the ureters are disconnected from the native bladder and may complicate future efforts to undivert the urine.

Cutaneous vesicostomy is a better option for temporary diversion of urine in a child with hostile bladder pressures. Vesicostomy drainage can easily be contained in a diaper in early childhood but is rarely a long-term option. Closure of the vesicostomy typically would be planned for early school-age when continence will become an issue for the child. This procedure would be combined with augmentation of bladder capacity and enhancement of bladder outlet resistance as indicated by urodynamic parameters. An abdominal wall stoma may be created to facilitate catheterization, and fecal incontinence refractory to maximal medical management could also be addressed during the same laparotomy with a MACE procedure

(Malone antegrade continence enema).

Obviously protection of the upper urinary tract takes precedence over continence, but better medical management and newer surgical techniques over the past several decades have facilitated both of these objectives and have virtually eliminated the need for long-term incontinent diversion. Institution of clean intermittent catheterization (CIC) and use of anticholinergic medications such as Oxybutinin (Ditropan) and Tolterodine (Detrol) to reduce bladder contractility and storage pressures and to increase the functional bladder capacity will provide upper tract stability and allow social continence in the majority of patients with neuropathic bladder.

Catheterization is well-accepted by children with impaired perineal sensation, particularly in those who have been catheterized since infancy. Catheterization per urethra may require transfers from the wheelchair to the toilet or may be difficult for those with limited dexterity, thereby limiting potential for self-care. Children with a sensate urethra and bladder neck, such as those with suboptimal bladder function after repair of exstrophy-epispadias complex, often tolerate CIC very poorly. Finally, adolescents in any of these groups are notoriously noncompliant with catheterization and any means by which their regimen may be simplified is beneficial.

In 1980 Paul Mitrofanoff, a French urologist, introduced a concept that significantly enhanced the management of children with congenital and acquired anomalies of the genitourinary tract. The classic Mitrofanoff procedure utilizes the appendix, which is brought up as a cutaneous stoma after being isolated on its vascular pedicle and implanted into the bladder. Similar to the natural mechanism and surgical technique to

prevent vesicoureteral reflux, the submucosal tunnel created within the bladder prevents leakage of urine through the channel (hence the term *continent catheterizable channel*). Ideally the stoma is camouflaged in the umbilicus but may require placement elsewhere on the abdominal wall.

In those children in whom the appendix is surgically absent or otherwise inadequate, the Mitrofanoff principle has since been applied to a variety of other tissues: ureter, bladder wall, intestine, stomach and even fallopian tube. Any of these tubes might be referred to casually as a Mitrofanoff, a catheterizable channel or a continent vesicostomy. In 1997 a South American surgery resident described a technique for reconfiguration of a short segment of ileum to create a very reliable catheterizable channel. The Monti ileovesicostomy, named for that resident, is now the most common appendiceal substitute and when it is used preferentially in urinary tract reconstruction, the appendix can be reserved for use as an *in situ* channel for administration of antegrade continence enemas. Use of a continent catheterizable channel is more convenient than catheterization per urethra in most patients but is particularly desirable in sensate patients and in wheelchair-bound females who otherwise must transfer to the toilet or a bed to catheterize.

Occasionally patients have difficulty using the channel and may create a false passage within the wall of the channel or just outside the bladder continence mechanism. The primary complication of continent catheterizable channels, however, is stomal stenosis. Critical stomal stenosis requiring surgical revision occurs in about 20% of classic Mitrofanoffs, 40% of continent bladder tubes and rarely in Monti ileovesicostomies. Although not clinically significant in all patients, bladder emptying is less efficient via

a catheterizable channel than through the urethra, largely due to difficulty emptying the dependent aspect of the bladder. In those patients prone to symptomatic urinary tract infection or bladder stones, routine irrigation of the bladder via a urethral catheter should be performed at least weekly.

In the patient with unsafe bladder storage pressures despite maximum anticholinergic therapy, bladder augmentation will be required to protect the upper tracts at the time of reconstruction for continence. Bladder volume is increased by bivalving the native bladder and adding on a generous detubularized segment of ileum or less commonly colon (*enterocystoplasty*, *ileocystoplasty* or *colocystoplasty*). The selected tissue is isolated from the GI tract and kept on a vascular pedicle. In patients with underlying renal insufficiency and acidosis, a wedge of stomach may be used instead or in composite with a segment of ileum.

The bowel segment maintains its absorptive and secretory properties and those features are responsible for the metabolic complications of bladder augmentation. For example, use of ileum or colon is associated with a hyperchloremic metabolic acidosis due to exchange of electrolytes across the bowel mucosa and due to absorption of ammonium and other acids from the urine. Reabsorption of ammonium from the urine disables the body's typical mechanism for handling an acute acid load. The resultant chronic acidosis is addressed by mobilization of bony buffers and may result in bone demineralization and linear growth abnormalities if untreated. The degree of metabolic derangement is proportional to the surface area of exposed mucosa and the time of contact with the urine. The management of other conditions can be confused by absorption of solutes from the urine. Dilantin is

reabsorbed across the bowel patch and the dose should be adjusted accordingly. Urine glucose dipsticks in diabetics may also be misleading due to absorption of glucose.

Stomach segments continue to secrete HCl, resulting in hypokalemic, hypochloremic metabolic alkalosis. A severe contraction alkalosis may result from an acute gastrointestinal illness. Cases refractory to salt supplementation and H₂ blockade may require a pump inhibitor or removal of the gastric patch. The acid load in the urine may result in hematuria and irritative symptoms in sensate patients. In composite augmentations, the electrolyte changes related to the ileum and stomach tend to neutralize each other. Off-the-shelf bioscaffold materials are currently being investigated as bladder substitutes to avoid these metabolic derangements.

The most concerning complication of bladder augmentation is perforation of the bowel segment. This is occasionally due to blunt trauma with a full bladder but most commonly is spontaneous. Although not proven, ischemia due to chronic overdistension is felt to play a role. Indeed most perforations seem to occur in adolescents and teens, who later give a history of delaying catheterization on the night of the perforation. Because many patients with augments have altered visceral sensation, they can become quite ill before there are obvious symptoms. Some cases have been fatal. Existing bacteriuria leads to peritonitis and sepsis. Patients with a ventriculoperitoneal shunt require externalization for weeks prior to replacement of the shunt. CT cystogram is the preferred diagnostic study.

Mucus production by the intestinal segment may increase the incidence of urinary tract infection and stones in the bladder. Daily bladder irrigation with saline will minimize these complications.

If a patient chooses not to use a channel it may stenose at the skin or bladder level over time, but failing to use the channel or losing the ability to access the channel is very serious when combined with a bladder augmentation and a bladder outlet procedure that precludes or limits emergency catheterization per urethra. Any child undergoing bladder augmentation and a bladder outlet procedure is committed to life-long catheterization, and failure to comply can be life-threatening. Removing the urethra as a pop-off valve for excess bladder volume and pressure can result in spontaneous perforation of the augmented bladder or damaging back-pressure to the upper urinary tract.

Our patients with bladder augmentations should be seen every six months for the first several years and then at least yearly to monitor electrolytes and to monitor the integrity of the upper tracts with ultrasound or IVP. Proper counseling of the patient and family to make sure that they understand the lifetime commitment is essential, but rebellion during the teenage years is common. Without incontinence as a reminder, the potential grave complications of failure to catheterize are forgotten. Reinforcement of these issues during clinic visits will be beneficial.



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