Learning Objectives
1. Describe a sacrococcygeal teratoma and the two distinct ways in which they generally present.
2. Explain the classification system used to categorize sacrococcygeal teratomas.
3. List two reasons post-operative follow up is needed for the first several years of life.

Clinical Presentation
• Sacrococcygeal teratomas generally present in two distinct fashions:
  - Newborns
    • Large predominately external masses found in utero or at the time of delivery. Rarely malignant.
  - Older Infants and Young Children
    • Presents with tumors confined to the pelvis.
    • Experience a much higher rate of malignancy.

Altman Classification of Sacrococcygeal Teratoma
• Type I tumors are predominately external (47%).
• Type II tumors present with both internal and external components (35%).
• Type III tumors are predominately hidden in the pelvis with very small external portion (9%).
• Type IV tumors are entirely hidden within the pelvis (10%).
Long Term Sequelae

• Long-term studies have demonstrated significant post-operative functional complications including constipation (30-36%) and urinary incontinence (19-22%).
• A neurogenic bladder has also been reported in 12% of children.

Post-Operative Follow Up

1. Constipation and urinary incontinence risks make it crucial for patients diagnosed with sacrococcygeal tumor to have very close outpatient monitoring during the first few years of life to ensure bowel and bladder function is normal as they go through toilet training.
2. A recurrence rate of 10-20% should be taken into consideration. More importantly, the average malignancy rate is around 50%, making post-operative follow up essential.
3. Follow up with Pediatric Surgery until age 5 is recommended. Alpha feta protein levels and physical exams including rectal exams to evaluate for mass are recommended routinely.

References and Acknowledgements

Acknowledgements
Drs. Deborah Billmire and Fredrick Rescorla for providing surgical images

References
Learning Objectives

- Define Sudden Unexpected Postnatal Collapse (SUPC)
- Understand the incidence of SUPC
- Identify risk factors for SUPC
- Able to identify signs of SUPC
- Able to state the implications for nursing practice to minimize SUPC

Sudden Unexpected Postnatal Collapse in the Healthy Newborn

- Characterized by apnea, bradycardia, cyanosis, limppness, and cardiorespiratory collapse in the health term newborn soon after birth
- A condition in which a previously vigorous, spontaneously breathing infant with a 5 minute Apgar score of 8 or greater unexpected becomes apneic and requires resuscitation
- Often during initial skin-to-skin contact or with first breastfeeding attempt
- Etiology is not completely understood, but most likely the result of accidental suffocation of the infant as a result of poor positioning, associated with maternal fatigue and/or distraction

What’s in a name?

- ALTE-apparent life-threatening event
- BRUE-brief, resolved, unexplained event
- SUPC-sudden unexpected postnatal collapse
- >37 weeks’ gestation at birth
- Appar score >8 at 5 minutes of life
- Collapse within 12 hours of birth in hospital
- Requires resuscitation after collapse with positive pressure ventilation
- Death or receives ongoing intensive care support

A Relatively New Clinical Phenomenon

Unintended Consequences of Current Breastfeeding, Skin to Skin, and Rooming In Initiatives

- The true incidence of SUPC is unknown because of the wide variation in the definitions of newborn events.
- Not uncommon that neonates who experience SUPC and respond favorably to resuscitation (near misses) are often not included in published documents.
- Given the lack of consensus and no International Classification of Diseases 10 (ICD-10) coding, it is very likely underreported.
- 2.6 to 133 cases per 100,000 births with the mortality rate of 0-1.1 per 100,000 live birth
- Median age at SUPC occurs at 70 minutes after birth, for infant without any underlying health concerns
- Median age with an underlying health concerns at SUPC is 195 minutes after birth
- Nearly one-third of the reported cases occur in the first 2 hours after birth (usually during the first breastfeeding attempt)
- Another one-third occur between 2 and 24 hours of life
- The final one-third occur between 1 and 7 days

Ferrarello & Carmichael, 2016; Rodriguez et al., 2017
Maternal Risk Factors for SUPC

- Primiparous status
- Maternal opiate analgesia
- General anesthesia within 8 hours of event
- Magnesium sulfate administration during labor
- Maternal body mass index > 25 kg/m²
- Large breasts
- Maternal fatigue/falling asleep during breastfeeding
- Prolonged labor
- Pain
- Sleep deprivation

Perinatal Risk Factors for SUPC

- Prenatal compromise
- Passage of meconium in utero
- Need for extensive neonatal resuscitation after delivery
- Delivery by cesarean birth
- Need for extensive repair after vaginal birth

Neonatal Risk Factors for SUPC

- Prone position of the infant while mother supine
- Infant fatigue
- Late preterm or preterm infant deemed safe to be left in the mother’s room
- Accidental suffocation due to occluded airway
- Underlying conditions
  - Cardiac disease-HLHS, interrupted aortic arch
  - Pulmonary disease-PPHN
  - Infection
  - Inborn error of metabolism
  - Airway-prolapsed epiglottis with laryngomalacia

Environmental/Situational Risk Factors for SUPC

- Breastfeeding-especially first attempt
- Unobserved skin-to-skin care with infant prone or side-lying on mother’s chest
- Minimal staff surveillance
- Fatigued parents
- Mother left alone with infant
- Parental distraction-Cell phone

Prevention of SUPC
Safely Keeping Mother and Baby Together

- Again, there is no doubt the benefits of early skin to skin contact, rooming in, and breastfeeding is undeniable!
- Balancing early skin-to-skin and promoting a safe environment for the mother baby dyad is crucial!

Interventions to Prevent SUPC

- Parent Education
  - Safe Skin to Skin Positioning
    - Mother is positioned with head of bed elevated
    - Infant is placed back in crib or with another person who is alert and awake
    - Proper positioning of the infant to ensure upper airway patency
    - Face can be seen and is in the sniffing position
    - Nose and mouth visible
    - Head is turned to one side
    - Neck is straight, not bent
    - Shoulders and chest are flat against mother; infant is covered up to the shoulders with blanket
    - Legs are fixed
  - General Education
    - Distraction-free breastfeeding and skin-to-skin
    - Re-emphasis of no distractions-no cell phone use
    - Education on maternal fatigue. It is common and when experienced, the infant should go back to the crib so that the mother does not risk falling asleep while holding the infant
Interventions to Prevent SUPC

- Staff Education
  - Encourage second clinical nurse at each delivery
  - Frequency of vital signs
  - Education
  - In-depth review of safe positioning
  - Mother in a reclined 35° to 80° position
  - Infant’s head, shoulders, and arms are elevated, making the infant’s face less likely to burrow into the breast
  - Simulation drills
  - Supporting parental education

- Surveillance Protocol
  - Pink and positioned
  - Respiratory, Activity, Perfusion, and Position Tool (RAPP)

Conclusion

- SUPC is a relatively new phenomenon that can lead to a devastating outcome for both the infant and the family
- It is crucial to identify risk factors that place the infant at a higher risk for SUPC
- Increased education to parents and staff, with more frequent surveillance can help to prevent SUPC
- Developing a charting template will increase compliance of more frequent surveillance

References


"Children who experience homelessness are among the most neglected and invisible of all."

Statistics

- 1 in 30 children
- 1 in 10 youth ages 18-24
- 2.5 million children each year are homeless

Youth Homelessness defined

- No fixed or regular nighttime residence
- Living in a public or non-private space
- Living in temporary shelter or living arrangement

Statistics...

- Nearly 2 million youth experience at least one night of homelessness
- 40% are LGBTQ and 40% have spent time in Foster Care
- 80% have experienced physical or sexual violence
What constitutes health:

10% Physical environment
20% Health care
30% Life behaviors
40% Socioeconomic factors

New City Initiative (Portland, OR) There are many factors that enter into homelessness, such as job loss, physical or mental disability, domestic violence, mental illness, drug and alcohol addiction, and others. But one important factor that is frequently overlooked is the breakdown of relationships and community that occurs when people become homeless. **People don't become homeless when they run out of money, at least not right away. They become homeless when they run out of relationships.**

Impacts of Homelessness

ACE STUDY and Homeless Youth

- Additional research on ACEs more specifically with homeless youth looked at the correlation between ACE scores and both Physician Trust Scale scores and Adult Attachment Scale scores.
- Interviewed youth between the ages of 16-24.
- Common themes identified:
  1) reported experiences of adult perpetrated trauma,
  2) suspicion of health care providers, and
  3) avoidance of health professionals.
- Results show that ACE scores are associated with lower trust in the medical profession, which is tied to less willingness to seek care, to share sensitive information, and to follow provider recommendations.

Implications for the Pediatric Focused APRN

- **BUILD TRUST**
  - Healthcare is usually complex – consider ACEs and disparities in healthcare and the physical and mental health needs can be overwhelming
  - Think MASLOW hierarchy of needs – food and shelter are a priority "Housing first" policy
  - Do as much as you can in a single visit when possible
  - Shelter life is not easy – often need to leave during the day, often no wifi, if late returning can be shut out
  - 504 plan for school can be helpful

Advocacy facts

- Social dollars have a net gain effect on the U.S. economy
- Cutting safety net programs greatly increases the impact for families and communities – greater mental health concerns, more urgent care and emergency room use and less preventative health care.
- Studies have shown that many antipoverty programs, especially those that target children, offer an excellent return on investment to taxpayers.
- Contrary to the myths - receiving benefits does not doom a person to a life of poverty.
**Things to encourage in self and others**

Show empathy

Stop and talk, or more importantly ask questions and listen

Give something tangible — a meal, a book, a blanket, socks, energy bars, hygiene kit etc. - ask what they need

Speak in resilient language — talk about strengths

Call for help if needed

---

**Summary:**

If you don’t ask you won’t know

Build trust — don’t make promises you can’t keep

Consider the complexities of being “healthy” and negotiate realistic goals

Foster relationship building in all youth

Be an advocate for homeless youth and help other youth become advocates in the community — share your stories and encourage youth to share theirs.

---

**Resources:**


Learning Objectives

• Describe the significance of penicillin allergy testing in the pediatric patient

What is the problem?

Prevalence

• Penicillin is most commonly reported drug allergy (32 million)
• Reported in approximately 10-20% hospitalized patients
• True prevalence is closer to 4-5% (some sources go as low as 1%)

What we know about penicillin allergy in children

• 5-10% of children will develop a rash while on amoxicillin
• A reaction to a penicillin during childhood infection is unlikely to be a true allergy
   95% of children who have had an amoxicillin-associated rash (MPE, hives) are not truly allergic to amoxicillin
• A penicillin allergy is not necessarily a life-long allergy
   Even if it was a true allergic reaction, 80% will tolerate amoxicillin after 10 years (50% after 5 years)

Take home message: we have over-diagnosis/unverified penicillin allergy labels that are negatively affecting patient outcomes

Why is it important?

From a patient safety standpoint:
• Increases hospitalization
• Increases morbidity
• C. diff, MRSA, VRE, surgical site infections

From a public health standpoint:
• Contributes to antibiotic resistance

From a financial standpoint:
• Increases cost of care

Fear of cross-reactivity of cephalosporins with penicillin often results in more use of broad-spectrum antibiotics

What can you do?

• If you have a patient that reports a penicillin allergy, ask questions
  • Dig deeper –
     What type of reaction? When did it happen? How was it treated?
     Do not list as an allergy if they have never taken it (i.e. family history only)
     Do not list a side effect (i.e. diarrhea) as an allergy

• Educate your families/patients
• Refer for testing
• Be a responsible prescriber
What is penicillin allergy testing?

- The gold standard is the Drug Provocation Test (DPT)
  - Give age-appropriate dose of amoxicillin and observe for 60 minutes
  - Rules out an IgE-mediated immediate reaction
- Occasionally skin testing is done before DPT
  - For anaphylaxis concerns
- Extended challenge
  - To identify a T-cell-mediated delayed hypersensitivity reaction
- Some research has also shown this to increase confidence with testing and increase likelihood of removing the allergy label and prescribing penicillin for future infections

Educational video explaining penicillin allergy

Can be found on YouTube – search for CCHMC PATS (Penicillin Allergy Testing Service)

Link: https://www.youtube.com/watch?v=cRBNTAOp2jw

References

Recognizing the Signs and Symptoms of Acute Disseminated Encephalomyelitis (ADEM) in Pediatric Primary Care

Paula Barbel, PhD, CPNP-PC

Disclosures
I have nothing to disclose

Learning Objectives
• To recognize the signs and symptoms of ADEM
• To describe the diagnostic criteria and imaging
• To describe the treatment and management of ADEM

Acute Disseminated Encephalomyelitis (ADEM)
• Definition: Rare acute monophasic immune-mediated inflammatory disorder of the central nervous system that results in demyelination and damage to the white matter of the brain (National Institute of Neurological Disorders and Stroke, 2019).
• Can occur after viral infections and rarely following vaccination and pneumococcal infections.
• The estimated rate is 1 in every 125,000 to 250,000 children.

Signs and Symptoms
• Fever
• Headache
• Nausea and vomiting
• Irritability
• Malaise/sleepiness
• Meningeal signs
• Ataxia
• Seizures and coma
• Altered LOC, speech disturbances, visual disturbances and focal neurological signs

Treatment
• Corticosteroids
• IVIG
• Plasmapheresis
Implications

- Although ADEM is rare, primary care providers must be able to recognize the signs and symptoms given that several other disorders such as multiple sclerosis and viral encephalitis may present with similar symptoms.

<table>
<thead>
<tr>
<th>ADEM versus MS</th>
<th>Clinically</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ADEM</strong></td>
<td><strong>MS</strong></td>
</tr>
<tr>
<td>Clinical</td>
<td>Multiphasic</td>
</tr>
<tr>
<td>Viral trigger</td>
<td>Unknown</td>
</tr>
<tr>
<td>CSF</td>
<td>Oblonginal bands</td>
</tr>
<tr>
<td>Spinal cord</td>
<td>Short segment lesions</td>
</tr>
<tr>
<td>Follow-up</td>
<td>New lesions</td>
</tr>
<tr>
<td>Monophasic</td>
<td></td>
</tr>
<tr>
<td>Often</td>
<td></td>
</tr>
<tr>
<td>Pilocytosis</td>
<td></td>
</tr>
</tbody>
</table>
What’s an ARM have to do with it?

Anal Rectal Manometry
Debra Browne, RNP, CPNP
Division of Gastroenterology, CHLA

Introduction
Used to differentiate several disease processes that may present with similar symptoms but require different treatments.

Examples: Hirschsprung’s, spinal cord lesions, neuromuscular disease, anal rectal pathology and dyssynergia defecation

ARM
• A = ANAL
• R = RECTAL
• M = MANOMETRY
• Used for diagnosing defecation disorders in constipated patients.

ARM
Measures:
--pressures of the anal sphincter muscles
--sensation in the rectum
--muscle and nerve reflexes that are needed for normal bowel movements

Test takes about 30 min. to complete

Description of the Study
• Catheter with sensors—flexible or rigid 3-D
• Covered with a latex free “condom” with a balloon that can be inflated.
• Connected to a computer with software that will read the study
Manometry computer

Manometry catheter

3-D Catheter

Patient Prep
• Empty rectum
• Left lateral side lying position, knees flexed and hips at a 90 degree angle.
• Test best performed awake, skeletal and striated muscles affected by sedation. Can test RAIR under sedation/anesthesia.

Elements of Study
• **Resting pressure**: 40-50 mm Hg
• **Rectal Anal Inhibitory Response** (RAIR); reflexive relaxation of Internal Anal Sphincter caused by stool presence and is simulated by rapid balloon fill during the procedure.
• **Slow Fill**: prolonged return to normal resting suggests spinal cord involvement

Elements of the Study
• **Sensation**= first 30-60 ml; urge 90-120 ml; discomfort close to urge
• **Squeeze**: 1.5 x resting; assess sphincter strength and tone (voluntarily tightens anal sphincter, be sure not using abd. muscles)
• **Bear Down** = simulated defecation (asses anorectal and pelvic floor pressure changes) ability is acquired about age 5-6 years 25 % relaxation
• **Balloon Expulsion**= inflated with 50 ml water — assesses ability to evacuate simulated stool. Confirms presence of dyssynergia.
Results

• Increased rectal sphincter pressure = outlet obstruction
• Decreased rectal sphincter pressure = fecal incontinence
•Absent RAIR= Hirschsprung’s, anal achalasia—rectal biopsy needed to confirm.

Results cont.

• Inability to squeeze = incontinence; myogenic or neurogenic problem
• Inability to relax = dyssynergia
• Unable to expel balloon = dyssynergia or lack of understanding how body works.

Summary: the results of the study:

Help the clinician to:
Identify the specific defecation problem
Develop the correct treatment plan
Advancements in Conservative Management of Idiopathic Scoliosis

Carrie Chan, CPNP
Manager of Advanced Practice
Pediatric Orthopedic Nurse Practitioner
Stanford Children’s Health
Assistant Clinical Professor
University of California, San Francisco School of Nursing

March 26, 2020

Learning Objectives

- Describe advancements in bracing options and technologies
- Discuss multimodal methods of optimizing conservative care of idiopathic scoliosis

Types of Braces

<table>
<thead>
<tr>
<th>Full-time</th>
<th>Night-time</th>
</tr>
</thead>
<tbody>
<tr>
<td>Boston</td>
<td>Rigo Cheneau</td>
</tr>
<tr>
<td>Providence</td>
<td>Charleston</td>
</tr>
</tbody>
</table>

Efficacy

- Rigid > night time + elastic
- Boston TLSO gold standard
- Rigo Cheneau
- Early evidence equal/better success rate

- More higher quality studies to come!

Technology and Future of Bracing

- Digital modeling
- 3D printing
- Smart braces
  - Compliance monitoring: hours, tension, pressure
  - Bluetooth capability & smartphone application

Low Dose Imaging
Physiotherapy Scoliosis Specific Exercises (PSSEs)

- Therapeutic exercises, asymmetric breathing techniques, re-education of the neuromuscular system to achieve passive and active correction of the curvatures and elongation of the spine
- Schroth, Dobosiewicz, side-shift, SEAS

Schroth Method

- Most widely studied and used PSSE approach
- Three-dimensional principles of correction throughout the day to change habitual postures

PSSEs Treatment Goals

- Mild scoliosis: Reduce progression of scoliosis and postpone/avoid bracing
- Moderate scoliosis: Adjunctive to bracing to increase efficacy in avoiding scoliosis progression
- Avoid secondary effects of bracing (spinal stiffness, muscle strength loss)
- Improve compliance with bracing

PSSEs: What’s Next?

- Low quality evidence from prospective controlled cohort studies that PSSEs can reduce brace prescription
- Poor quality evidence of significant improvement vs. bracing alone
- Little data on patient centered outcomes (quality of life, back pain, psychological, cosmetic issues)
- BUT increasing patient interest
- Need more studies clarifying effectiveness

Dietary Supplementation

- Multiple studies have shown Vitamin D lower in AIS group
  - Positively correlated with calcium
  - Negatively correlated with Cobb angle
- Lam et al. (2017)
  - Placebo 46.7% progressed
  - 600 mg Ca + 400 IU VitD 24.4% progressed
  - 600 mg Ca + 800 IU VitD 21.7% progressed
  - Indicates daily supplementation 600mg Ca + 400/800 IU VitD3

Mehta Casting

- Serial casting for early onset scoliosis
  - Primary Goal: Resolution of deformity
  - Secondary Goal: Delay surgical intervention
  - Congenital curves: Did not correct, delay surgery 26 months
  - Best response if <2 years, idiopathic, <60°
  - Technique
    - Change every 2 months
    - Stop when curve <10-15° or when not getting lasting correction/improvement
    - Consider transition to brace
Thank you!

Carrie Chan, CPNP

cachan@stanfordchildrens.org

Manager of Advanced Practice
Pediatric Orthopedic Nurse Practitioner
Stanford Children’s Health

Assistant Clinical Professor
University of California, San Francisco School of Nursing
New Pediatric Drugs for Primary Care

Teri Moser Woo PhD, ARNP, CPNP-PC, FAANP
Professor and Director of Nursing
Saint Martin's University

Disclosures
No Disclosures

Learning Objectives
- Review newly FDA approved pediatric medications

New Cystic Fibrosis Drugs
- Trikafta (elexacaftor/ivacaftor/tezacaftor)
  - Approved for patients 12 years and older with cystic fibrosis who have at least one F508del mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene
  - Increases predicted percent predicted forced expiratory volume in one second (ppFEV1)
  - CFTR gene is estimated to be present in 90% of the cystic fibrosis population

Sickle Cell Disease
- Oxbryta (voxelotor)
  - Approved for age 12 yrs and older
  - Inhibitor of deoxygenated sickle hemoglobin polymerization
  - Sickle cells are less likely to bind together and form the sickle shape
  - Accelerated approval – further trials ongoing
Duchenne muscular dystrophy

- Vyondys 53 (golodirsen) injection
  - Treats patients with Duchenne muscular dystrophy who have a confirmed mutation of the dystrophin gene that is amenable to exon 53 skipping
  - Vyondys 53 masks exon 53 in the mRNA of the DMD gene so the protein synthesis machinery can skip this exon and piece together the remaining exons to make a smaller, but functional, dystrophin protein.
  - Accelerated approval – further trials ongoing

Expanded Age Range in Pediatric Patients

- Mavyret (glecaprevir and pibrentasvir) for chronic hepatitis C genotype 1, 2, 3, 4, 5 or 6 infection
  - Expanded the indication to adolescents 12 years and older or weighing at least 45 kilograms (kg); previously approved in adults
- Solvadi (sofosbuvir) for chronic hepatitis C genotype 1 or 4 infection
  - Expanded age range to patients age 3 to 11 years
- Harvoni (ledipasvir and sofosbuvir) for chronic hepatitis C genotype 1, 4, 5 or 6 infection
  - Expanded age range to patients age 3 to 11 years; and at least 17 kg
- Welchol Chewable Bar (colesevelam HCl)
  - Reduce LDL-C levels in boys and postmenarchal girls, 10 to 17 years of age, with heterozygous familial hypercholesterolemia as monotherapy or in combination with a statin after failing an adequate trial of diet therapy

References