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Acute Care Case Studies

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Speaker Disclosure

- No financial disclosures
- Off-label utilization of medications is discussed in Case #3
- The intention of the third case in this presentation is to describe principles regarding pain management rather than specific doses or regimen recommendations. All providers are responsible for consulting institution-approved references when recommending or prescribing medication regimens.

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Learning Objectives

- Analyze key concepts in the care of acute or critically ill pediatric patients through an interactive case study discussion
- Recognize associated co-morbidities in the patient with trisomy 21, including current guidelines related to screening
- Understand sequalae of single ventricle physiology and atrioventricular valve regurgitation
- Describe a stepwise approach to intractable pain that includes pharmacologic and nonpharmacologic interventions

3

Question

- What is your primary area of practice?
  A. Emergency department
  B. Acute care/subspecialty
  C. Critical Care
  D. Primary Care

4

Question

- How many years have you practiced as an Advanced Practice Provider?
  A. Student
  B. <5 years
  C. 5-10 years
  D. >10 years

5

Case #1

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Case 1 Presentation

Chief Complaint:

9-year-old-female with history of trisomy 21 and abnormal neurologic exam who presents with abnormal MRI findings after a scheduled outpatient brain MRI this morning

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Trisomy 21

- According to the Center for Disease, Down Syndrome is the most frequent chromosomal disorder in the United States (CDC, 2023)
- The estimated prevalence in the US was 1 in 700 live births
- Outcomes for children with Down Syndrome can be positively impacted across their lifespan based on awareness of important issues impacting this patient population during childhood and adolescents

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Trisomy 21

Common Physical Characteristics:

- Hypotonia
- Small, brachycephalic head
- Epicanthal folds
- Flat nasal bridge
- Upward-slanting palpebral fissures
- Brushfield spots
- Small nose/mouth
- Deep plantar groove
- Cognitive impairment (various degree)

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Trisomy 21

TABLE 1 Medical Problems Common in Down Syndrome

Condition	%
Hearing problems	75
Vision problems	60–80
Nystagmus	3–33
Glaucoma	<1–7
Nasolacrimal duct occlusion	3–36
Cataracts	3
Strabismus	36
Refractive errors	36–80
Keratoconus	1–13
Obstructive sleep apnea	55–79
Otitis media with effusion	50–70
Congenital heart disease	40–50
Feeding difficulty	31–80
Respiratory infection	20–36
Dermatologic problems	56
Hypodontia and delayed dental eruption	20
Congenital hypothyroidism	2–7
Antithyroid antibody positive (Hashimoto thyroiditis; incidence dependent on age)	13–39
Hyperthyroidism	0.65–3
Thyroid disease by adulthood	50
Gastrointestinal atresias	12
Seizures	1–13
Hematologic problems	
Anemia	1.2
Iron deficiency	6.7
Transient abnormal myelopoiesis	10
Leukemia	1
Autoimmune conditions	
Hashimoto thyroiditis	13–39
Graves' disease	1
Celiac disease	1–5
Type 1 diabetes	1
Juvenile idiopathic arthritis	<1
Alopecia	5
Symptomatic atlantoaxial instability	1–2
Autism	7–19
Hirschsprung disease	<1
Moyamoya disease	Down syndrome 26 times greater in patients with Moyamoya than Down syndrome in true births

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Trisomy 21: Abbreviated AAP Guidelines:

	Birth- 1 Year	1-5 Years	5-12 years	12-21 years
Exam	-Assess for myelopathic S/SX, any developmental regression	-Assess for myelopathic S/SX, any developmental regression	-Assess for myelopathic S/SX, any developmental regression	-Assess for myelopathic S/SX, any developmental regression
Screening	-Obtain History -6 months: hearing/vision, assess for sleep, disorder breathing	-Vision annually, sleep all visits -If S/SX of Myelopathy: obtain c-spine plain films	-Vision biennially, sleep all visits -If S/SX of Myelopathy: obtain c-spine plain films	-Vision Optho sets, sleep all visits -If S/SX of Myelopathy: obtain c-spine plain films
Anticipatory Guidance	-Cervical Spine -S/SX Myelopathy	-Cervical Spine -S/SX Myelopathy (Biennially)		

Signs and symptoms of myelopathy: asymmetry of movement, weakness, and, on examination, increased deep tendon reflexes.

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Trisomy 21

AAP Guidelines: Summary of Down Syndrome-Specific Care

Supplemental Information

SUPPLEMENTAL FIGURE 1. Summary of Down syndrome-specific care.

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Case 1 Continued: Chart Review

7 years prior to admission:

3-year-old patient presents to (spine clinic) today for initial evaluation of C1-C2 subluxation, the patient was referred by the pediatrician.

- Parents report that an x-ray was performed, and they were sent here for further evaluation
- Birth and developmental history is significant for trisomy 21 and a leg discrepancy
- Parents deny and cervical spine symptoms –no pain in the arms or neck
- The patient is non-verbal. She can walk and move all extremities spontaneously with out difficulty.
- Parents deny radiculopathy, paresthesias, subjective weakness, any significant neurologic deficits, myelopathy, and bowel or bladder dysfunction. Denies exacerbating or relieving factors. Current treatments: None"

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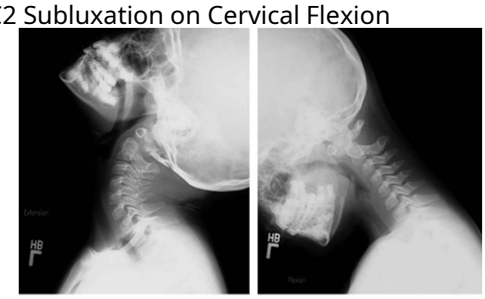
C1-C2 Subluxation on Cervical Flexion



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Atlantoaxial Instability

- C1-C2 instability
- Approximately 10-30%
- Exact mechanisms not fully understood: Multifactorial

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Question

Considering the AAP guidelines and our patient story, what is your recommendation for follow up after the spine clinic appointment?

A. Follow up spine clinic appointment in 2 years with ongoing monitoring by primary care provider
B. Urgent Neurology consult
C. Outpatient MRI
D. Direct admission to the children's hospital

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Case 1 Continued: Chart Review

2 years prior to admission:

- Sleep Study obtained
- ENT Office
- Hospital Admission:
  - ENT H&P:
    - 7-year-old female presents with OSA for planned T&A
    - PMH: DS, history of PFO-closed, no hypothyroidism
    - PLAN: to OR for T&A, admit for observation, 4 years ago – 2mm increase in subluxation on C1-C2 with flexion vs extension. See by Spine surgeon, repeat neck X-ray and possible consult if needed.
  - Anesthesia H&P:
    - OSA, hypertrophic adenoids/tonsils, developmental delay, hypotonia, Syndromes: "has negative genetic syndromes"
    - Case: orally intubated: 5.0 cuffed ETT, secured at 14 CM, Mac 2 blade, Grade 2 view, 2 attempts (redundant tissue- stylet on 2nd attempt) technique Atraumatic, cricoid pressure, easy bag/mask
    - Extubated discharged POD #1

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Question

Based on the facts pulled out from the chart review, what concerns do you have with the documentation for the patient admitted for a T&A?

A. Insufficient pre-anesthesia documentation of trisomy 21
B. Insufficient documentation of positioning / precautions taken
C. Absent parent report: cervical spine precautions
D. Lack of updated imaging
E. All of the above

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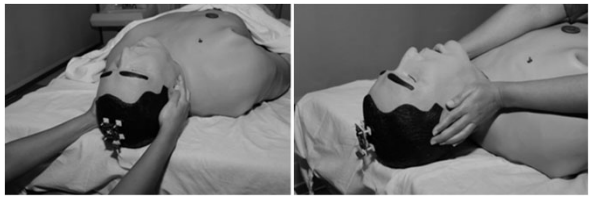
Immobilization Techniques:



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Case Overview

3-year-old female
- Trisomy 21
- Asymptomatic with C1-C2 2 mm subluxation on c-spine X-ray

7-year-old female
- T&A

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Case 1 Continued: Chart Review

1 year prior to admission:

Orthopedic Surgery Office visit:

- "8-year-old female with Down Syndrome presents today with concern regarding left lower extremity shorter than her right extremity with difficulty ambulating"
- BLE x-ray obtained
- Recommend follow up with PT

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Chart Review

6 months prior to admission:

Orthopedic Surgery Office visit:

- 9-year-old female with down syndrome and pes planovalgus foot deformities with bilateral SMO braces that are fabricated by an outside orthopedics company
- Documented Travel: No Physical Therapy
- Exam:
  - Left foot more rigid than right
  - decreased subtalar range of motion increased tone on left,
  - passive ankle dorsiflexion neutral to the knee with knee bent in flexion on the left
  - clonus BLE
  - While standing planovalgus foot deformity exaggerated significant pronation while ambulating
  - Weakness noted while ambulating and after several steps has to hold on to parent
  - Questionable ataxia and posturing of the upper extremities
- Referral to Ped Neurology

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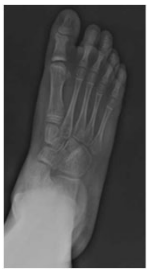
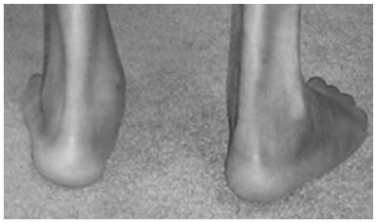
Plangovalgus: Foot Deformity





Kadhim (2014)

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Case Overview

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Case 1: Chart Review

3 months prior to admission:

- Ortho follow up appointment
- Neurology consult
  - "Gait is markedly abnormal. Prominent inversion and external rotation of her ankles while walking she tends to drag her feet while walking, she does not appear to be able to dorsiflex. She tends to "W" sit"

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Question

You are the provider in the specialty clinic: What is your recommendation for the BEST next step?

1) Direct admission to the children's hospital
2) Instruct the family to follow up with ortho and primary care provider
3) Follow up in neurology clinic in 1 month
4) Urgent Imaging: MRI

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Case 1: Chart Review

Day of admission:

- Sedation consult: MRI
- MRI brain and lumbar spine

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Question

You are a part of the sedation team, what are you considering/documenting prior to sedating this patient:

A. NPO status
B. History of trisomy 21
C. History of C1-C2 subluxation
D. Developmental regression
E. All of the above

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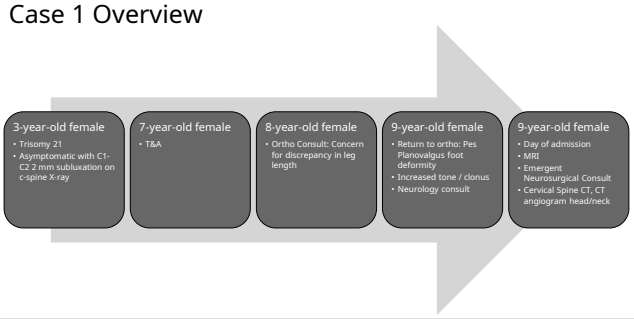
Case 1 Overview



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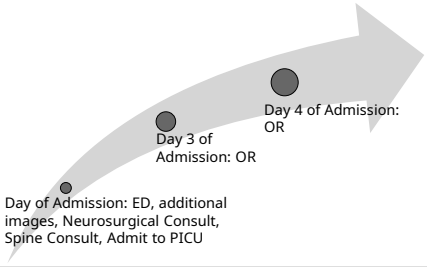
Case 1 Continued



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Acute Care Management: Spinal Cord Injury

- Inpatient care: multifactorial
  - Positioning
  - Intubation/mechanical ventilation/respiratory failure
  - Neurogenic shock
  - Cardiovascular management
  - Autonomic nervous system management
  - Neurogenic bowel/bladder
  - Pharmacologic management
  - Rehabilitation
- Multidisciplinary team approach
- Family involvement / education

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Acute Care Management: Spinal Cord Injury

CLINICAL INVESTIGATIONS

Optimal Timing of Tracheostomy in Injured Adolescents*

OBJECTIVES: To evaluate the optimal timing of tracheostomy for injured adolescents.

DESIGN: Retrospective cohort study.

SETTING: Trauma facilities in the United States.

PATIENTS: Adolescents (age 12–17 yr) in the National Trauma Data Bank (2007–2016) who were ventilated for greater than 24 hours and survived to discharge.

Elissa K. Butler, MD[1]
Elizabeth Y. Killen, MD, MPH[2]
Jonathan I. Groner, MD[3]
Saman Arbabi, MD, MPH[4]
Monica S. Vavilala, MD[5]
Frederick P. Rivara, MD, MPH[2]

Pediatric Spinal Cord Injury: Recognition of Injury and Initial Resuscitation, in Hospital Management, and Coordination of Care

Kyle Lemley[1] Paul Bauer[2]

ADVANCED JOURNAL OF EMERGENCY MEDICINE, 2020;4(2):e28 *Benmelouka et al*

Review Article DOI: 10.22114/ajem.v0i0.256

A Review on the Etiology and Management of Pediatric Traumatic Spinal Cord Injuries

Amira Benmelouka[1], Laila Salah Shamseldin[2], Anas Zakarya Nourelden[3], Ahmed Negida[4,5,6*]

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Early Mobility

- Bundled, interdisciplinary interventions
  - Improved adult ICU outcomes
- PICU Up! study ongoing

://

Azamfrei et al(2023)

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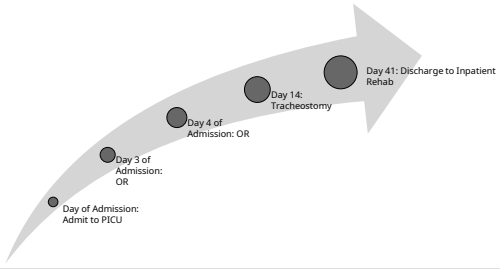
Case 1 Continued



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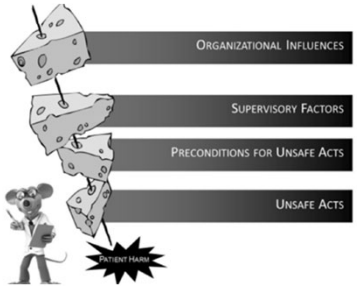
Patient Safety

The Swiss Cheese Model



Wiegmann et al (2022)

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Case 1: Summary

- Primary Care
  - AAP guidelines Down Syndrome: monitoring / screening
  - Family involvement
  - Clear Overt Documentation
- Acute Care
  - Management of spinal cord injury: acute / chronic
  - Multidisciplinary team
  - Early involvement support services
  - Family involvement
  - Clear Overt Documentation

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References

Appireddy R, Ranjan M, Durafourt BA, Riva-Cambrin J, Hader WJ, Adelson PD. Surgery for Moyamoya Disease in Children. Journal of Child Neurology. 2019;34(9):517-529. doi:10.1177/0883073819844854

Azamfirei, R., Mennie, C., Dinglas, V.D. *et al.* Impact of a multifaceted early mobility intervention for critically ill children — the PICU Up! trial: study protocol for a multicenter stepped-wedge cluster randomized controlled trial. *Trials* **24**, 191 (2023). https://doi.org/10.1186/s13063-023-07206-2**/doi-org.echo.louisville.edu/10.3171/2023.3.PEDS22353**

Bauer, Jennifer M. MD, MS*; Dhaliwal, Virkamal K. BS, MHS†; Browd, Samuel R. MD, PhD‡; Krengel, Walter F. III MD*. Repeat Pediatric Trisomy 21 Radiographic Exam: Does Atlantoaxial Instability Develop Over Time?. Journal of Pediatric Orthopaedics 41(8):p e646-e650, September 2021. | DOI: 10.1097/BPO.0000000000001884

Benmelouka A, Shamseldin LS, Nourelden AZ, Negida A. A Review on the Etiology and Management of Pediatric Traumatic Spinal Cord Injuries. Adv J Emerg Med. 2019 Oct 10;4(2):e28. doi: 10.22114/ajem.v0i0.256. PMID: 32322796; PMCID: PMC7163256.

Bouaré F, Lmejjati M, Mpando D. Spontaneous atlanto-axial dislocation and trisomy 21: causal factors and management. Pan Afr Med J. 2019 May 6;33:3. doi: 10.11604/pamj.2019.33.3.18109. PMID: 31303948; PMCID: PMC6607459.

Bull, M.J., Trotter, T., Santoro, S., Christensen, C. & Grout, R. (2022). Health supervision for children and adolescents with down syndrome. *Pediatrics, 149* (5). https://doi.org/10.1542/peds.2022-057010

Butler, Elissa K. MD1–3; Killien, Elizabeth Y. MD, MPH1,4; Groner, Jonathan I. MD5; Arbabi, Saman MD, MPH1,2; Vavilala, Monica S. MD1,6; Rivara, Frederick P. MD, MPH1,7. Optimal Timing of Tracheostomy in Injured Adolescents*. Pediatric Critical Care Medicine 22(7):p 629-641, July 2021. | DOI: 10.1097/PCC.0000000000002681

Center for Disease Control and Prevention (2023, October 10) Facts about down syndrome. https://www.cdc.gov/ncbddd/birthdefects/downsyndrome.html#print

De Jesus, Clarines, Garcia Pena, Barbara, MD, MPH, Lozano, Juan, Manuel MD, MSc & Maniaci, Vincenzo. (2015). Cervical Spine Motion During Airway Management Using Two Manual In-line Immobilization Techniques: A Human Simulator Model Study. Pediatric Emergency Care, 31, 627-632. https://doi.org/10.1097/PEC.0000000000000245

Graber, T. J., Baskin, P. L., Soria, C., Greenberg, M., Gabriel, R. A., Brzenski, A. (2021). An assessment of perioperative respiratory adverse events and difficult intubation in pediatric patients with trisomy 21.. Paediatric Anaesthesia, , 31(4), 410-418. https://dx.doi.org/10.1111/pan.14138

Hedequist, Daniel & Mo, Andrew. (2020). Os Odontoideum in Children. Journal of the American Academy of Orthopaedic Surgeons, 28, e100-e107. https://doi.org/10.5435/JAAOS-D-18-00637

Isaacs, A. M., Narapareddy, A., Nam, A., Hutcheson, K., Stone, M., & Bonfield, C. M. (2023). Surgical treatment of craniovertebral junction instability in children with Down syndrome: a systematic review. *Journal of Neurosurgery: Pediatrics*, *32*(2), 163-172. htt

Kadhim, Muayad; Miller, Freeman. Pes planovalgus deformity in children with cerebral palsy: review article. Journal of Pediatric Orthopaedics B 23(5):p 400-405, September 2014. | DOI: 10.1097/BPB.0000000000000073

Lemley K, Bauer P. Pediatric Spinal Cord Injury: Recognition of Injury and Initial Resuscitation, in Hospital Management, and Coordination of Care. J Pediatr Intensive Care. 2015 Mar;4(1):27-34. doi: 10.1055/s-0035-1554986. PMID: 31110847; PMCID: PMC6513171.

Macdonald M, Crawford J, Datta AN. Children With Trisomy 21 and Lennox-Gastaut Syndrome With Predominant Myoclonic Seizures. Journal of Child Neurology. 2021;36(11):1027-1033. doi:10.1177/08830738211026072

Wiegmann DA, Wood LJ, Cohen TN, Shappell SA. Understanding the "Swiss Cheese Model" and Its Application to Patient Safety. J Patient Saf. 2022 Mar 1;18(2):119-123. doi: 10.1097/PTS.0000000000000810. PMID: 33852542; PMCID: PMC8514562. Understanding the "Swiss Cheese Model" and Its Application to Patient Safety (nih.gov)

Case #2

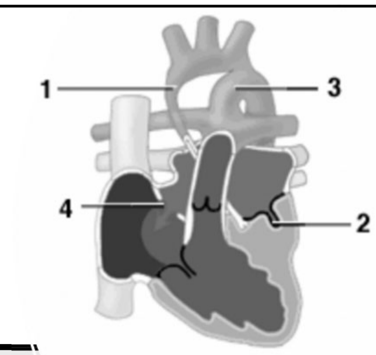
Case 2

4-month-old with complex medical history including trisomy 21, Hypoplastic Left Heart Syndrome (HLHS) and complete tracheal rings

- s/p Norwood procedure with Blalock-Taussig (BT) shunt
- s/p Slide tracheoplasty
- Atrioventricular valve regurgitation (AVVR)
- Abnormal lung parenchyma
- Narcotic dependence
- s/p Gastrostomy tube placement
- Not a candidate for Glenn procedure

Hypoplastic Left Heart Syndrome

- Functionally univentricular heart
- Subtypes
  - Mitral and aortic stenosis
  - Mitral and aortic atresia
  - Mitral stenosis and aortic atresia
  - Aortic stenosis and mitral atresia
  - Intact atrial septum



Jacobs (2022)

https://www.cincinnatichildrens.org/patients/child/encyclopedia/defects/graphicsummaries/hlhsGS

Question

- What is the approximate prevalence of HLHS among all congenital heart disease?
  - A. 2%
  - B. 5%
  - C. 10%
  - D. 20%

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Question

- What is the approximate rate of mortality among congenital heart disease?
  - A. 2%
  - B. 5%
  - C. 10%
  - D. 20%

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Epidemiology

- 2.3 per 10,000 live births
- Underestimated overall occurrence
- Males > females
- 10% of all congenital heart deaths
- Post-operative mortality of 16%
- 8-12% interstage mortality rate

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Question

- If you are at a center that performs stage 1 palliation for HLHS, what is the primary surgical procedure performed?
  - A. Hybrid procedure
  - B. Norwood with BT shunt
  - C. Norwood with Sano shunt
  - D. Uncertain

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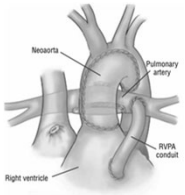
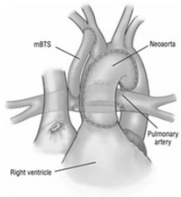
Norwood Procedure

Sano Shunt



Blalock-Taussig Shunt



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The NEW ENGLAND JOURNAL of MEDICINE

ORIGINAL ARTICLE

Comparison of Shunt Types in the Norwood Procedure for Single-Ventricle Lesions

Richard G. Ohye, M.D., Lynn A. Sleeper, Sc.D., Lynn Mahony, M.D., Jane W. Newburger, M.D., M.P.H., Gail D. Pearson, M.D., Sc.D., Minmin Lu, M.S., Caren S. Goldberg, M.D., Sarah Tabbutt, M.D., Ph.D., Peter C. Frommelt, M.D., Nancy S. Ghanayem, M.D., Peter C. Laussen, M.B., B.S., John F. Rhodes, M.D., Alan B. Lewis, M.D., Seema Mital, M.D., Chitra Ravishankar, M.D., Ismee A. Williams, M.D., Carolyn Dunbar-Masterson, B.S.N., R.N., Andrew M. Atz, M.D., Steven Colan, M.D., L. LuAnn Minich, M.D., Christian Pizarro, M.D., Kirk R. Kanter, M.D., James Jaggers, M.D., Jeffrey P. Jacobs, M.D., Catherine Dent Krawczeski, M.D., Nancy Pike, R.N., Ph.D., Brian W. McCrindle, M.D., M.P.H., Lisa Virzi, R.N., M.S., M.B.A., and J. William Gaynor, M.D., for the Pediatric Heart Network Investigators

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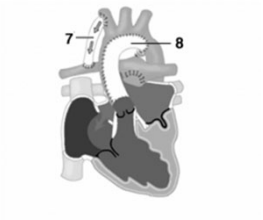
Goals of Norwood Procedure

- Unobstructed systemic blood flow
- Unobstructed pulmonary venous return
- Restrict pulmonary blood flow



https://www.cincinnatichildrens.org/patients/child/encyclopedia/defects/graphicsummaries/hlhsGS

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$$\frac{Qp}{Qs} = \frac{Ao\ O_2\ sat - MV\ O_2\ sat}{PV\ O_2\ sat - PA\ O_2\ sat}$$

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Atrioventricular Valve Regurgitation

- Tricuspid valve
- Abnormal valve morphology
- Volume load on single ventricle
- Ventricular dysfunction
- Significant mortality risk

Lee et al, (2012)

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Risk Factor Analysis for Second-Stage Palliation of Single Ventricle Anatomy

Timothy M. Lee, BA, Ranjit Aiyagari, MD, Jennifer C. Hirsch, MD, Richard G. Ohye, MD, Edward L. Bove, MD, and Eric J. Devaney, MD

Division of Pediatric Cardiovascular Surgery, Section of Cardiac Surgery, and Division of Pediatric Cardiology, University of Michigan Medical School, Ann Arbor, Michigan

Combined ventricular dysfunction and atrioventricular valve regurgitation after the Norwood procedure are associated with attrition prior to superior cavopulmonary connection

Sheri L. Balsara, MD, MS,[a] Danielle Burstein, MD, MSCE,[a,b] Richard F. Ittenbach, PhD,[c] Michelle Kaplinski, MD,[d] Monique M. Gardner, MD,[e] Chitra Ravishankar, MD,[a] Joseph Rossano, MD,[a] David J. Goldberg, MD,[a] Marlene Mahle, RN,[a] Matthew J. O'Connor, MD,[a] Christopher E. Mascio, MD,[a] J. William Gaynor, MD,[a] and Tamar J. Preminger, MD[a]

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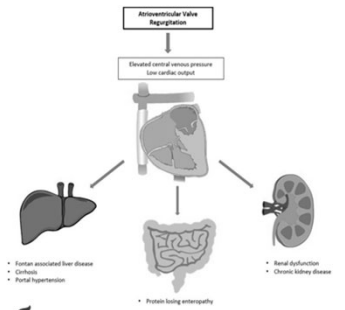
Sequelae:

- Ventricular failure
- End organ dysfunction
- Shock



Stephanie Y. Tseng. Journal of the American Heart Association. Atrioventricular Valve Regurgitation in Single Ventricle Heart Disease: A Common Finding Associated With Increased Detection and Mortality. Volume: 9, Issue: 11, DOI: (10.1161/JAHA.119.012727)

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Case 2: Update

- Severe atrioventricular valve regurgitation following Norwood with BT shunt, despite valve repair
- Lung parenchyma is abnormal and biopsy was inconclusive
- Prolonged course of mechanical ventilation
- Extubated, required significant oxygen

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Question

- What do you recommend as next steps for this patient?
  A. Proceed with Bidirectional Glenn procedure
  B. Additional valve repair
  C. Plan to list for heart transplant
  D. Change goals of care to comfort

53

Question

- How would you feed this child at home?
  A. Nasogastric tube
  B. Nasojejunal tube
  C. Gastrostomy tube
  D. Total Parenteral Nutrition

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THE JOURNAL OF PEDIATRICS • www.jpeds.com ORIGINAL ARTICLES

Variation in Feeding Practices following the Norwood Procedure

Linda M. Lambert, MSN-FNP[1], Nancy A. Pike, PhD, CPNP-AC[2], Barbara Medoff-Cooper, RN, PhD, FAAN[3], Victor Zak, PhD[4], Victoria L. Pemberton, RN, MS[5], Lisa Young-Borkowski, MSN, RN[6], Martha L. Clabby, MD[7], Kathryn N. Nelson, DNP, CPNP[8], Richard G. Ohye, MD[9], Bethany Trainor, RN, BSN[10], Karen Uzark, PhD, CPNP[11], Nancy Rudd, CPNP-AC/PC[12], Louise Bannister, RD[13], Rosalind Korsin, RN, BSN[14], David S. Cooper, MD, MPH[15], Christian Pizarro, MD[16], Sinai C. Zyblewski, MD[17], Bronwyn H. Bartle, MSN-CPNP[18], and Richard V. Williams, MD[19], for the Pediatric Heart Network Investigators*

55

Pediatr Surg Int (2016) 32:285–289
DOI 10.1007/s00383-015-3854-1

ORIGINAL ARTICLE

Feeding gastrostomy in children with complex heart disease: when is a fundoplication indicated?

Jennifer L. Carpenter[1] · Timothy A. Soeken[1] · Alfred J. Correa[1] · Irving J. Zamora[1] · Sara C. Fallon[1] · Mark J. Kissler[1] · Charles D. Fraser Jr[2] · David E. Wesson[1]

Results:
- study reviewed open procedure
- minor complications
- minimal major complications
- few needed fundoplication
- 29% had HLHS

Complications:
- shunt occlusion
- dislodgement
- infection
- bleeding

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JOURNAL FOR SPECIALISTS IN PEDIATRIC NURSING
the international evidence-based practice journal for nurses caring for children and families

ORIGINAL ARTICLE

Parents' perceptions during the transition to home for their child with a congenital heart defect: How can we support families of children with hypoplastic left heart syndrome?

Sarita March

First published: 18 June 2017 | https://doi.org/10.1111/jspn.12185 | Citations: 21

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Case 2: Update

- Patient returned to the ICU from open gastrostomy tube placement 4 hours ago. Patient remains endotracheally intubated and NPO.
- Called to the bedside for crying, agitation, and "looking bad".
- Exam:
  - Cold, mottled extremities
  - Ventilator 50% FiO2, Rate 30, Vt 8mL/kg, PIP 35-38, PEEP +6
- Vitals:
  - HR 175, RR 54, Sat 74%, BP 125/85 MAP 95, temp 36.8C
- ABG: 7.25/38/40/16/-8/lactate 6

58

Question

- What do you think is going on with your patient?
  A. Septic shock
  B. Developing pneumonia
  C. Inadequate afterload reduction
  D. Inadequate pain control

59

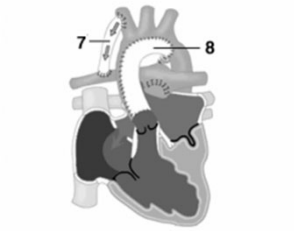
Question

- The nurse administers PRN dose of Fentanyl. The patient calms down, however the exam and vital signs are relatively unchanged. What is your next intervention?
  A. More Fentanyl
  B. Start Milrinone infusion
  C. Start Nicardipine infusion
  D. Administer diuresis
  E. Administer antibiotics

60

Norwood

- Balance pulmonary and systemic blood flow
- ↑ SVR = ↓ oxygen delivery
- ↑ SVR = ↑ RV volume
- Unbalanced Qp:Qs
- Augment SVR



https://www.cincinnatichildrens.org/patients/child/encyclopedia/defects/graphicsummaries/hlhsGS

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Management of AVVR

Augment afterload

- Milrinone
- Nicardipine
- Nitroprusside
- Sedative
- Positive pressure ventilation

Augment Preload

- Diuresis

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Pediatric Critical Care Medicine
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CARDIAC INTENSIVE CARE SUBSCRIBED AHEAD OF PRINT

Nicardipine or Nitroprusside for Postoperative Blood Pressure Control in Infants After Surgery for Congenital Heart Disease: Single-Center Retrospective Noninferiority and Cost Analysis, 2016–2020

Wong, Rudolph J. MD, MBA[1]; Mruk, Allison L. PharmD, BCPPS[2]; Grimaldi, Lisa M. MD[3]; Patel, Reena CPNP-AC[4]; Mirea, Lucia PhD[5]; Engelhardt, Kevin P. MD* Author Information

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Case 2: Update

- Patient was started on Milrinone and Nicardipine infusions with some improvement in BP, which was followed by improvement in perfusion. Patient was sedated and paralyzed.
  - Vitals:
    - HR 150, RR 45, Sat 75%, BP 100/72 MAP 82, temp 36.8C
  - ABG:
    - 7.33/38/42/18/-4/lactate 3.5

64

Case 2: Update

- Perfusion worsened and developed anuria
- NIRs 50s→20s
- ABG: 7.22/34/37/16/-9/lactate 7
- BUN 55, Creatinine 0.9
- PIP 39-40

- Acidosis progressed and lead to cardiac arrest
- CPR was brief and parents elected to stop compressions

65

Case 2: Summary

- Uncontrolled afterload
- Ventricular dysfunction
- Multiorgan failure
- Shock
- Gastrostomy tube?
- Pain control?
- Withdrawal?
- Earlier afterload reduction?

66

References

- Beke, D.M. (2016). Norwood procedure for palliation of hypoplastic left heart syndrome: Right ventricle to pulmonary artery conduit versus Blalock-Taussig shunt. *Critical Care Nurse, 36(6).*
- Carpenter, J.L. et al (2016). Feeding gastrostomy in children with complex heart disease: when is fundoplication indicated? *Pediatric Surgery International, 32,* 285-289.
- Jacobs, J. (2022). Hypoplastic left heart syndrome: Definition, morphology, and classification. *World Journal of Pediatric and Congenital Heart Surgery, 13(5),* p559-554.
- Kumar, K.R. et al (2019). Cardiovascular physiology for intensivists. In R.M. Ungerleider, J.N. Meliones, K.N. McMillan, D.S. Cooper, J.P. Jacobs (Eds.), *Critical Heart Disease in Infants and Children (pp111-133).* Elsevier.
- Lambert, L.M. et al (2014). Variation in feeding practices following the Norwood procedure. *The Journal of Pediatrics, 164(2).*
- Lee, T.M. et al (2012). Risk factor analysis for second stage palliation of single ventricle anatomy. *Annals of Thoracic Surgery, 93,* p614-619.
- March, S. (2017). Parents' perceptions during the transition to home for their child with a congenital heart defect: How can we support families of children with hypoplastic left heart syndrome? *Journal for Specialist in Pediatric Nursing*. DOI: 10.1111/jspn.12185
- Ohye, R.G. et al (2010). Comparison of shunt types in Norwood procedure for single-ventricle lesions. *New England Journal of Medicine, 362*(21).
- Quintessenza, J. et al (2019). Hypoplastic left heart syndrome. In R.M. Ungerleider, J.N. Meliones, K.N. McMillan, D.S. Cooper, J.P. Jacobs (Eds.), *Critical Heart Disease in Infants and Children (pp778-795).* Elsevier.
- Tseng, S.Y. (2020). Atrioventricular valve regurgitation in single ventricle heart disease: A common problem associated with progressive deterioration and mortality. *Journal of the American Heart Association, 9(11),* e015737
- Wong, R.J. et al. (2024). Nicardipine or nipride for postoperative blood pressure control in infant after surgery for congenital heart disease: single center retrospective noninferiority and cost analysis 2016-2020. *Pediatric Critical Care Medicine, 25(6).*

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Case #3

68

Question

- Does your institution have a palliative care service?
  A. Yes – with 24/7 access to providers
  B. Yes – with limited access to providers
  C. No

69

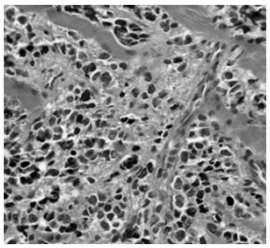
Case 3: Patient "C"

- 20-year-old non-binary young adult
- Presented with lower back pain and symptoms consistent with sciatica to urgent care and the emergency department
- Treated with muscle relaxers, NSAIDS, advised to stretch

70

Case 3

- Neurosurgery and interventional radiology consulted for biopsy
- Neurosurgery performed decompressive L4 laminectomy due to neurologic changes
- Pathology: metastatic alveolar rhabdomyosarcoma



https://imagebank.hematology.org/image/60887/alveolar-rhabdomyosarcoma-alveolar-rhabdomyosarcoma-forearm-mass

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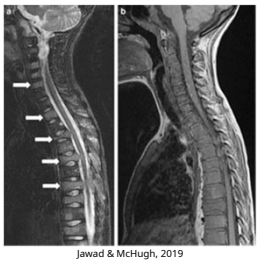
Alveolar Rhabdomyosarcoma

- Malignant tumor of mesenchymal cell origin
- Third most common extracranial solid tumor in children
- Majority of cases occur spontaneously, some association with certain familial syndromes
- Primary site of origin: head/neck (35-40%), genitourinary tract (<25%), extremity (20%), truncal primary/misc. site (~10% each)
- Between 15-25% of new diagnoses have distant metastasis

72

Alveolar Rhabdomyosarcoma

- Diagnosis:
  - Complete history
  - Physical exam
  - Laboratory evaluation
  - +/- Bone marrow aspiration/biopsy
  - PET scan
  - +/- Nuclear medicine bone scan
  - CT scan
  - MRI



Jawad & McHugh, 2019

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Alveolar Rhabdomyosarcoma

- Staged according to spread, disease involvement
- Prognosis based on stage, surgical resectability, site, size, age, histology, presence of metastasis, and regional lymph node involvement
- Treatment modalities include surgical resection, radiation therapy, and systemic, combination chemotherapy
- Challenges associated with treatment in the setting of recurrent disease

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Case 3

- Treated per ARST2031
- Vincristine, Actinomycin, Cyclophosphamide (VAC) therapy
- Complicated by significant nausea and vomiting
- Persistent Pain
  - Mixed type: neuropathic, nociceptive

75

Types of Pain

Neuropathic: caused by a lesion or disease of the somatosensory nervous system

Nociceptive: detection of potentially damaging external stimuli by sensory neuron input to the CNS

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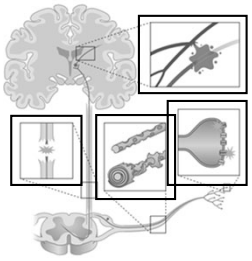
Pain Pathophysiology

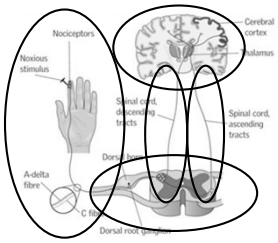
- Neuropathic Pain
  - Peripheral receptors
  - Peripheral nerves
  - Central nervous system
  - Brain lesions



77

Pain Pathophysiology

- Nociception
  - Peripheral mechanisms
  - Spinal cord
  - Spinocortical pathways
  - Cortical detection
  - Descending pathways



https://dm3omg1n1n7zx.cloudfront.net/rcni/static/journals/ns/25/3/ns2010.09.25.3.35.c7989/graphic/ns_v25_n3_7989_0002.jpg

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Approach to Pediatric Pain

- Evaluation with history and physical exam
- Treat underlying causes
- Utilize integrative therapies and interprofessional expertise
- Basic analgesics, opioids
- Consider adjuvants

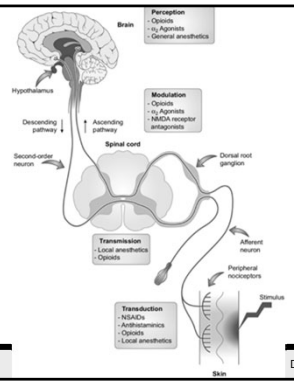
79

Question

- Which of the following analgesics may offer relief of both types of pain this patient is experiencing?
  A. Morphine
  B. Gabapentin
  C. Ketorolac
  D. Methadone

80

Pharmacologic Interventions



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Pain Interventions

- Basic analgesia: acetaminophen
- Opioids: oxycodone, hydromorphone PCA
- Topical: lidocaine patch
- Adjuvant: gabapentin

82

Question

- This patient weighs 65 kg and is experiencing severe pain. When the opioid was rotated to hydromorphone, the initial dose was:
  Hydromorphone 2.6 mg PO every 4 hours
- You want to prescribe an additional dose for breakthrough pain and select:
  A. Hydromorphone 1 mg PO every 2 hours PRN breakthrough pain
  B. Hydromorphone 3 mg PO every 4 hours PRN breakthrough pain
  C. Hydromorphone 1.6 mg PO every 2 hours PRN breakthrough pain
  D. Hydromorphone 1 mg PO every 4 hours PRN breakthrough pain

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Case 3

- Significant, refractory pain despite interventions
- Required increasing PCA settings in the absence of side effects
- Elected to transition to methadone for baseline pain control
- Hydromorphone utilized for breakthrough pain

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Pain Interventions

- Methadone:
  - Stereoisomer acts as NMDA receptor antagonist
    - Prevents opioid tolerance and potentiates opioid effects
    - Indication for use in neuropathic pain syndromes
  - Opioid receptor activity works on mu, delta, kappa
  - Lipophilic, rapid GI absorption, and onset of action
  - No active metabolites, no biotransformation required
  - Hepatic metabolism
  - No dose adjustment in renal failure
  - Extended half-life
  - Risk for prolonged QT interval

85

Pain Interventions

- Methadone
  - Potency varies with exposure to other opioids
  - Several strategies for conversion from other opioids using daily oral morphine equivalents
  - Complex process that can be dangerous, recommend consultation with pain or palliative specialists

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Methadone Conversion

Daily oral morphine dose equivalents methadone	Conversion ratio of oral morphine to oral
<100 mg	3:1 (i.e. 3 mg morphine = 1 mg
101 – 300 mg	
301 – 600 mg	
601 – 800 mg	
801 – 1000 mg	
>1001 mg	20:1

Due to incomplete cross-tolerance, it is recommended that the initial dose is 50-75% of the equianalgesic dose.

This is just one example of a conversion table to serve as an example, not a dose recommendation

https://www.mypcnow.org/fast-fact/methadone-for-the-treatment-of-pain/

87

Case 3: Ongoing Palliative Care

- Followed across inpatient and outpatient settings
- Symptom management: disease-related pain, nausea, vomiting, mucositis
- Build rapport, support decision-making
- Ongoing discussion about goals of care
- Check-ins to ensure goals and care received align

88

Case 3: Four Months Later

- New/worsening back pain
- MRI and PET obtained:
  - Metastatic disease within the mediastinum
  - Metastasis in the left common femoral node
  - FDG-avid lesions:
    - Right scapula, manubrium/sternal body, T4, T9, T10, T12, L2-L5, S2 vertebral bodies, bilateral iliac bones, left proximal and distal femur, posterior crural region
- Treatment regimen changed to Protocol 0921 and palliative radiation initiated

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Pain Regimen

- Opioids:
  - Methadone 10 mg by mouth BID
  - Oxycodone 5-10 mg by mouth every 6 hours PRN
- Adjuvants:
  - Dexamethasone 4 mg by mouth
  - Duloxetine 60 mg daily
  - Gabapentin 800 mg by mouth TID

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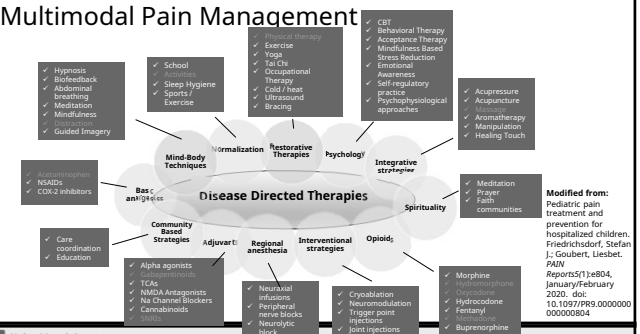
Multimodal Pain Management



Modified from: Pediatric pain treatment and prevention for hospitalized children. Friedrichsdorf, Stefan J.; Goubert, Liesbet. *PAIN Reports*5(1):e804, January/February 2020. doi: 10.1097/PR9.0000000000000804

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Case 3: Symptom Management

- Complaints of cramping muscle pain in extremity receiving radiation
  - Relieved by massage, physical therapy, time
  - Other pain well controlled on previously described regimen
- Radiation-related nausea
  - Regimen includes granisteron, olanzapine, aprepitant, scopolamine patch
  - PRN regimen: lorazepam, promethazine, hydroxyzine
- Opioid-induced constipation
  - Docusate, polyethylene glycol 3350

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Case 3: Two Months Later

- Presented to the ED with worsening pain
- Disease progression noted on CT, MRI, PET
- Received hydromorphone during the admission for acute pain
- Interventional radiology / anesthesia consulted for spinal root block
- Enrolled in clinical trial at outside hospital

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Case 3: Six Weeks Later

- Presented to the emergency department with uncontrolled lower back pain
- Weakness with ambulation, “feet have been cold”
- Intact bladder and bowel function, no change in paresthesia
- Worsening pain requiring increased PRN oxycodone
- Significant constipation

94

Case 3

- Admit to inpatient unit
- Pain regimen:
  - Hydromorphone, intermittent doses escalated to PCA
  - Lidocaine patch
  - Increase methadone
  - Continue gabapentin, dexamethasone, duloxetine
- Pain not controlled; palliative contacted for additional PCA recommendations

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Case 3: Palliative Care Plan

- Dilaudid PCA:
  - Continuous: 0.5 mg/hr → 0.75 mg/hr
  - Demand: 0.25 mg → 0.5 mg → 0.75 mg
  - Lockout: 15 minutes → 10 minutes (bolus helpful but not lasting long enough)
  - Increases of 50-100% as patient was tolerating without respiratory depression and still experiencing severe pain
- Continue to increase methadone based on opioid requirement
- Bowel regimen: Polyethylene glycol 3350 BID, Senna QHS, methylnaltrexone
- Canine therapy, music therapy

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Approach to Pediatric Pain

- Evaluation with history and physical exam
- Treat underlying causes
- Utilize integrative therapies and interprofessional expertise
- Basic analgesics, opioids
- Consider adjuvants
  - Gabapentinoids
  - Tricyclic antidepressants
  - Alpha-2 adrenergic agonists
  - NMDA-receptor-channel blocker
  - Sodium channel blockers

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Approach to Refractory Pain

- Consider non-pharmacodynamic factors
  - Is the disease progressing?
  - Are other therapies effective?
  - Is the medication being absorbed appropriately?
  - Could drug interactions be contributing to efficacy?
  - What is the patient's renal function?
  - What role are pharmacogenetics playing?
  - Is the patient experiencing psychological distress?
  - What is the cognitive status?
  - Could anxiety or delirium be contributing to the pain experience?

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Question

- What adjuvant would you recommend, especially given the concern for opioid-refractory pain?
  A. Ketamine
  B. Lidocaine
  C. Dexmedetomidine
  D. Amitriptyline

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Case 3

- Worsening pain despite escalating opioid therapy, concern this is becoming opioid-refractory
- Adjuvants attempted
  - Ketamine
  - Dexmedetomidine

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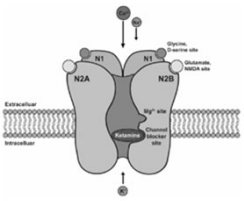
Pain Interventions and Targets

Off Label Use

- Ketamine
  - NMDA-receptor antagonist
  - Interacts with nicotinic, muscarinic, and opioid receptors
  - Subanesthetic doses can be effective for neuropathic and acute pain
  - Limited risk of respiratory or cardiovascular depression
  - Adverse effects rare at low doses
    - Intracranial hypertension, tachycardia, psychotomimetic phenomena



https://wou.edu/chemistry/chemistry-degree/student-activities-2/chemistry-corner/ketamine/

Chou, et al (2020); Friedrichsdorf & Postier (2019); Prommer (2019)

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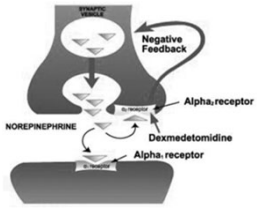
Pain Interventions and Targets

Off Label Use

- Dexmedetomidine
  - Alpha-2-adrenergic receptor agonist
  - Sedation, anxiolysis, and analgesia
  - Effective for neuropathic and nociceptive pain
  - Respiratory function often preserved
  - Dose-limiting hypotension and bradycardia may develop



https://www.openanesthesia.org/keywords/epidural-adjuvants-clonidine-and-dexmedetomidine/

Friedrichsdorf & Postier (2019); Mahmoud et al (2020)

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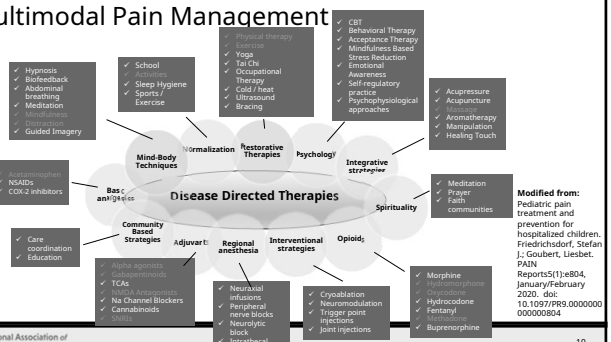
Multimodal Pain Management



Modified from: Pediatric pain treatment and prevention for hospitalized children. Friedrichsdorf, Stefan J.; Goubert, Liesbet. PAIN Reports5(1):e804, January/February 2020. doi: 10.1097/PR9.0000000000000804

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Question

- What adjuvant therapy do you have the most experience with at your institution?
  - A. Lidocaine
  - B. Dexmedetomidine
  - C. Ketamine
  - D. None of the above

104

Question

- Does your institution have policies to support continuous infusions of adjuvants (ketamine, dexmedetomidine, etc) outside of the intensive care unit?
  - Yes
  - Yes, but only for end-of-life situations
  - No

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Case 3

- Pain initially managed in the PICU, transitioned to oncology unit
  - Ketamine: developed a "loopy", dysphoric feeling, dose reduced to 2 mcg/kg/min
  - Dexmedetomidine: developed confusion, infusion discontinued
  - Continued to require an aggressive bowel regimen
  - Hydromorphone PCA continued, able to wean slightly
  - Methadone doses increased
  - Gabapentin doses optimized
- Acceptable pain management (3/10) reported
- MRI obtained, consistent with continued disease progression

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Case 3

- Salvage chemotherapy initiated
- Continuing to wean PCA as no demands were required as increased methadone and adjuvants were utilized
- Continued to prioritize being as alert as possible with tolerable pain
- Identified metastatic disease on head CT, hip radiograph
- Transition goals to comfort-focused plan of care at home

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Case 3 Summary

- Longitudinal role of palliative care across disease trajectory
- Step-wise approach to pain management
- Utility of adjuvant therapies in the setting of opioid refractory pain
- Alignment of goals and care delivered, honoring patient preference for alert mental status over complete pain control

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References

- Chour, C. Z., Hopkins, T. J., Badiola, I., Gazelka, H. M., Gohil, S., Kitt, A., Luke, W. R., Vaughan, C. L., Jones, C. A., & Varhabhatla, N. (2020). Top ten tips palliative care clinicians should know about interventional pain and procedures. *Journal of Palliative Medicine, 23*(10). 1386-1391.
- Dinakar, P., & Stillman, A. M. (2016). Pathogenesis of pain. *Seminars in Pediatric Neurology, 23*(3), 201-208.
- Dureja, G. P., Iyer, R. N., Das, G., Ahdal, J., & Narang, P. (2017). Evidence and consensus recommendations for the pharmacological management of pain in India. *Journal of Pain Research, 10,* 709-736.
- Finnerup, N. B., Kuner, R., & Jensen, T. S. (2021). Neuropathic pain: From mechanisms to treatment. *Physiological Reviews, 101*, 259-301.
- Friedrichsdorf, S. J., & Goubert, L. (2020). Pediatric pain treatment and prevention for hospitalized children. *PAIN Reports,* doi:10.1097/PR9.0000000000000804
- Friedrichsdorf, S. J., & Postier, A. C. (2019). Recent advances in pain treatment for children with serious illness. *Pain Management, 9*(6), 583-596.
- Gazelle, G., & Fine, P. G. (2019) Methadone for the treatment of pain. Mypcnow.org/fast-fact/methadone-for-the-treatment-of-pain/
- Jayakar, S., Shim, J., Jo, S., Bean, B. P., Singec, I., Woolf, C. J. (2021). Developing nociceptor-selective treatments for acute and chronic pain. Science Translational Medicine, 13 (619). DOI: 10.1126/scitranslmed.abj9837
- Jawad, N., & McHugh, K. (2019). The clinical and radiologic features of pediatric rhabdomyosarcoma. *Pediatric Radiology, 49*, 1516-1523.
- Linardic, C. M., & Wexler, L. H. (2021). Rhabdomyosarcoma. In S. M. Blaney, P. C. Adamson, & L. J. Helman (Ed.). *Pizzo and Poplack's pediatric oncology* (8th ed). Wolters Kluwer.
- Mahmoud, M., Barbi, E., & Mason, K. P. (2020). Dexmedetomidine: What's new for pediatrics? A narrative review. *Journal of Clinical Medicine, 9*(9), DOI: 10.3390jcm9092724
- Prommer, E. (2019). Ketamine in palliative care. https://www.mypcnow.org/fast-fact/ketamine-in-palliative-care/

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Questions?

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