AMERICAN COLLEGE OF EMERGENCY PHYSICIANS
SCIENTIFIC ASSEMBLY
1999

The Critically Ill Neonate

The severely ill neonate is a challenging problem, especially in the era of early-discharge newborns. The lecturer will review the critical findings on physical examination, outline essential and cost-effective diagnostic studies, and discuss initial indicated therapies.

- Develop a differential diagnosis for the critically ill neonate, especially for early-discharge newborns.
- Discuss proper neonatal stabilization and resuscitation.
- Identify clinically diagnostic findings on the physical and laboratory examinations of the neonate.

TH-197
Thursday, October 14, 1999
8:00 AM - 9:55 AM
Room # N212
Las Vegas Convention Center

FACULTY

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THE CRITICALLY ILL NEONATE

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NEWBORN DELIVERY and RESUSCITATION IN THE ED

PREPARATION PRIOR TO DELIVERY

- Have neonatal resuscitation kit or cart available - to include both airway and vascular access equipment
  - include small charts with drug dosages inside
- Warming bed
- Hotline to nearby NICU
- Non-sterile gloves in your back pocket

- Quickie questions for Mom:
  - due date – are you dealing with a premature birth?
  - only one fetus inside uterus – are you dealing with multiple births?
  - gestational diabetes - are you dealing with a potentially large newborn that will be more difficult to deliver?
- Take a look at Mom's underwear – might be able to tell about meconium – yes or no, thick or thin?

DELIVERY OF NEWBORN

- Fortunately, usually not a problem !
- If possible, suction nose then mouth before newborn is delivered
  - too vigorous stimulation of posterior pharynx can cause bradycardia
  - if meconium is present, attempt suction with 10F or larger suction (usually not possible because newborn delivers too quickly)
- Prevent newborn from hitting the ground!

MECONIUM – WHAT IS IT?

- Meconium is the newborn's first bowel movement while still in-utero; usually a sign of distress!
- Meconium aspirated into the newborn's lungs can cause a pneumonitis with permanent sequelae

- Whether or not ALL meconium, watery or thick, needs to be suction out of the TRACHEA remains controversial
- All newborns, with watery or thick meconium, do need their nose then mouth suction when the head is delivered

- Most experts agree, if thick or particulate meconium present or newborn is "depressed",
  DO NOT STIMULATE THE NEWBORN AFTER DELIVERY!
- *Thick meconium or depressed newborn* needs to have trachea suctioned with an ET tube attached to a meconium aspirator attached to wall suction (wall suction should be set to < 100 mm Hg)
  - controversial as whether or not to suction the active, vigorous newborn with meconium
- The ET tube is the SUCTION CATHETER! Do not attempt to pass a small suction catheter *through* the ET tube; the size of any suction catheter *through the ET tube* will be *too small* for removal of meconium
  - alternative to the ET tube is to use a large (size 12 F) suction catheter directly into the trachea
- How long to suction trachea? Generally, not longer than 3-5 seconds at a time.
- If newborn is severely depressed (bradycardic or dropping heart rate), then positive-pressure ventilation may need to be applied even if some meconium still exists in the airway
- Can use the same ET tube, if necessary, to deliver positive-pressure ventilation

RESUSCITATION BASICS – in order !

- Suction nose then mouth
- Dry 'em off then piss 'em off !! (after meconium suctioning, if necessary)
- Blow-by O2
- Bag-and-mask ventilation (positive-pressure)
- Intubate
- Chest compressions
- Epinephrine

- Intervention, then re-evaluate!
- KEEP NEWBORN WARM!

NORMAL FETAL CARDIOPULMONARY PHYSIOLOGY TRANSITION

- During fetal life, lungs/alveoli are fluid-filled sacs and blood flow through the lungs is markedly diminished
- Majority of blood flow during fetal life is diverted away from the lungs via the ductus arteriosus (connects pulmonary artery to the aorta)
- When newborn takes first breath at birth, the lungs expand with air and fetal lung fluid gradually leaves the alveoli (process of labor may only facilitate the process of removal of lung fluid; majority of the fluid is absorbed through the lymphatics and blood vessels within the lungs)
- Arterioles open and allow more blood flow to the lungs
- Ductus arteriosus begins to close soon after the first breath; functionally closed usually by 24 hours but can take up to several days to close

- First several breaths a newborn takes may require two to three times the negative pressure required for any succeeding breaths, i.e. IT TAKES EFFORT! Anything that inhibits the newborn's ability to generate this pressure can result in respiratory depression!

REASONS FOR HYPOXEMIA AT BIRTH

- Problems clearing fetal lung fluid – fluid in alveoli is not cleared quickly; usually due to newborn not making adequate respiratory efforts; if newborn has not taken an initial breath, assume that no expansion of alveoli has occurred and the alveoli remain filled with fluid
- Persistent fetal circulation – arteriolar vasoconstriction in the lungs persists after birth process

REASONS FOR INADEQUATE RESPIRATORY EFFORTS (LEADING TO DECREASED CLEARANCE OF LUNG FLUID)

- Mom's age > 35 or < 16yo
- Maternal diabetes or hypertension
- Maternal hemorrhage
- Prematurity or multiple fetuses
- Drugs given to mom or taken by mom prior to birth
- Congenital neuromuscular diseases
- Congenital malformations
- Abnormal presentation causing hypoxia
- Meconium-stained amniotic fluid

PHYSIOLOGY OF APNEA

- Neonate deprived of oxygen will have a brief period of rapid breathing ➜ respiratory movements soon stop, heart rate falls, tone becomes floppy, and neonate soon develops *primary apnea*
- During *primary apnea*, stimulation and oxygen will usually induce spontaneous respirations
- If apnea continues, neonate soon develops deep gasping respirations ➜ heart rate continues to fall, blood pressure falls, and neonate is now flaccid ➜ gasping soon stops and neonate develops *secondary apnea*
- During *secondary apnea*, blood pressure and heart rate continue to fall until neonate develops cardiac arrest

• During *secondary apnea*, neonate is unresponsive to stimulation and will not resume spontaneous respiratory efforts ➜ resuscitation must now include assisted ventilation, oxygen, and potentially cardiac compressions or medications
• Neonate born apneic may already be in *primary* or *secondary apnea;* these two conditions are virtually indistinguishable from each other; infant is not breathing and the heart rate < 100 bpm

• A newborn in *primary apnea* will usually respond to simple stimulation and oxygen; CAUTION – respiratory effort that does resume may be irregular or may be ineffective!
• A newborn in *secondary apnea* will not resume spontaneous breathing on his or her own; assisted ventilation with positive-pressure ventilation with oxygen usually necessary

• ASSUME NEWBORN IS IN *SECONDARY APNEA* UNTIL PROVEN OTHERWISE!
• DELAY IN INITIATING ASSISTED VENTILATION CAN RESULT IN LONG DELAY IN ESTABLISHING SPONTANEOUS RESPIRATIONS !

PERSISTENT FETAL CIRCULATION

• Pulmonary vessels remain constricted in the newborn, leading to hypoxemia and acidosis.
• In the presence of hypoxemia and acidosis, the newborn's pulmonary vessels remain even more constricted ➜ persistent fetal circulation
• Persistent fetal circulation may be improved by increasing oxygenation (ventilating with 100% oxygen) and if severe, may require correction of acidosis with bicarbonate (controversial)

BAGGING OF NEONATES

• Initiated in newborns who are apneic or whose heart rate is < 100bpm
  - measure newborns heart rate at cardiac apex or brachial artery or umbilical stump
• Occiput is large so place towel under the shoulders, approximately 1 inch
• Avoid hyperextension of the neck as this may actually close off the newborn's airway
• Choose a mask that fits – bridge of the nose to cleft of the chin, without pushing on the eyes
• Squeeze/Release/Release at a rate of 40 – 60 breaths per minute
• Less compliant lungs means higher pressures needed to generate air movement into the lungs
• If self-inflating bag has a pop-off valve, may need to depress it while ventilating infant
• Aim for chest rise only – entire tidal volume is only 25cc
• If pressure gauge available,
  - initial few breaths after delivery (requiring more pressure) 30-40 cm H2O
  - normal lungs soon after delivery 15-20 cm H2O
• Nasal airways are avoided due to large adenoids and size of nares
• Oral airways work well - measure level of gums to angle of the jaw

INTUBATING NEONATES

• Size 3.5 – 4.0 ETT, uncuffed tube for term babies > 3000g or > 38 wks
  - size 3.5 for neonate 2000 – 3000 g or 34-38 wks
  - size 3.0 for neonate 1000 – 2000 g or 28-34 wks
  - size 2.5 for a premature neonate < 1000 g
  - tapered ET tubes may be more difficult to place correctly into trachea since view is obstructed by the wide part of the tube
• Size 0 or 1 straight laryngoscope blade
• Don't need paralytics generally when intubating neonates
• Watch for reflex bradycardia associated with the laryngoscope
• Finesse is the name of the game !!!
• Tongues are larger, epiglottis is longer, cords are more anterior
• Place towel under the shoulders to align airway
• Breath sounds normally heard over the stomach because of transmission of sounds; listen for gurgling
• Don't pull a good tube!!
• Pediatric CO2 detectors now available

- Cut off any extra tube extending beyond 4 cm from neonate's lips – this will reduce the amount of dead space

CARDIAC COMPRESSIONS

- Immediately necessary for any newborn whose heart rate is < 60 bpm after 15-30 seconds of good ventilation, or if the heart rate is between 60-80 and not increasing
- 2 finger method - 1 finger breadth below nipple line, compress down ½ inch – ¾ inch
- hand wrap method - wrap 2 hands around the newborn's chest, use thumbs to do compressions
- Rate: 120 per minute, interposed with ventilations (3:1)

INTRAVENOUS ACCESS

- Umbilical line - use a 3.5 or 5F umbilical catheter or feeding tube and insert into vein
  - normal umbilicus has 2 arteries and 1 vein (like Mr.Bill from *Saturday Night Live)*
  - insert catheter just below skin level and free blood flow is present (use pickups or small clamp to pull umbilical cord straight for easier catheter insertion)
  - if a young neonate (< 7days) returns to ED in extremis, can still attempt umbilical line if crusted scab is still present; difficult if mom's using alcohol religiously
- Jugulars are hard since neonates have no neck
- Prep groin well if attempting femoral vein catheter; avoids infection in the joint

MEDICATIONS

- Epinephrine
  - indicated in the resuscitation when adequate ventilation and cardiac compressions fails to increase heart rate > 60 - 80
  - 1:10,000 concentration used in neonates
  - dose is 0.01 – 0.03 mg / kg (0.1 – 0.3 ml / kg) via IV, umbilical line, or ET tube
  - consider using higher dose (0.1 – 0.2 mg / kg = 1 – 2 ml / kg) if using ET tube or neonate does not respond to first dose
  - in order to deliver small amounts of epinephrine down the ET tube, may need to dilute with normal saline to deliver at least 2 ml of volume

- Volume expanders
  - indicated in the resuscitation when there is evidence or suspicion of acute blood loss with signs of hypovolemia
    - neonate can lose 10-15% blood volume and only show mild decrease in blood pressure
    - > 20% loss can result in pallor, weak pulses but good heart rate, poor response to resuscitation, decreased blood pressure
  - whole blood (O-negative crossmatched with mother's blood)
  - 5% albumin-saline solution
  - normal saline
  - Ringer's lactate
  - dose for all volume expanders = 10 ml / kg IV or umbilical given over 5 minutes

- Sodium bicarbonate
  - indicated in the resuscitation when there is prolonged arrest that does not respond to other therapy
  - 0.5 mEq / ml = 4.2% solution is the concentration used in neonates
  - dose = 2 mEq / kg IV or umbilical given slowly over 2 minutes

- Naloxone (Narcan)
  - indicated in the resuscitation when there is severe respiratory depression and a history of maternal narcotic administration within the past 4 hours
  - 0.4 mg / ml or 1.0 mg / ml solution is the concentration used in neonates
  - dose = 0.1 mg / kg IV, umbilical, or ET

- CAUTION – if mom is narcotic-addicted, naloxone may induce withdrawal seizures in the newborn

- Glucose
  - indicated for evidence of hypoglycemia – acceptable glucose in neonates > 40
  - D 10W concentration used in neonates
  - dose = 0.5 – 1.0 g / kg (5 – 10 ml / kg) IV or umbilical

LAB TESTS

- Heel sticks are great for a glucose and hct
  - Check Dextrose early in neonates; hypoglycemia can be end result of many different processes
  - Normal hct is 55%; low 30's nadir by 2 months of age
  - WBC 7 - 28,000 during first month of life
- IV's and labs: Prep the neonate for a blood culture every time you stick him for blood
- 24 or 26 gauge catheters
- Use antecubital area, hands, scalp veins
- Difficult to get an arterial stick in neonates
  - Capillary blood gas can give the pH and the CO2 ---> if normal, great !!

NOT YOUR TYPICAL NEWBORN:

- Choanal atresia – congenital blockage of one or both of the posterior nares by a membrane or bone
  - newborns are obligate nose breathers except when crying
  - newborn will be pink and oxygenated when crying and then become apneic and cyanotic when quiet – will usually be apparent during the first few minutes of life
  - an oral airway or ET tube must be inserted and left in place until surgery to correct defect
- Pierre-Robin Syndrome – congenital abnormality that results in an abnormally small mandible; can be very difficult to bag-and-mask ventilate and requires an oral airway
- Diaphragmatic hernia – suspected if newborn has scaphoid abdomen and difficulty breathing
  - intubate early since bag-and-mask ventilation will only allow more air to enter the bowel and compromise lung expansion
- Omphalocoele
- Spina bifida
- Premature – if < 1000 grams, majority will require endotracheal intubation

NORMAL NEONATAL ANATOMY AND PHYSIOLOGY

WEIGHTS AND GROWTHS

- most term infants will lose weight (10%) during first few days but level off by 5 - 7 days old
- most term infants will regain their birth weight by 10 - 14 days and then gain 20 - 30 grams (~1oz)/day for 1 - 2 months

NORMAL PULMONARY FUNCTION

- normal respiratory rate is 30 - 40 per minute
- neonates are obligate nose-breathers ---> watch that nasal cannulas don't cause respiratory distress !
- infant's ribs are aligned horizontally, so in order to increase thoracic diameter to inhale, infant must lower his diaphragm (if stomach is distended with air, diaphragm will not function properly and child will not be able to ventilate well ---> use an NG or OG tube when trying to ventilate a critically ill infant)

PERIODIC BREATHING PATTERN

- occurs in normal full term infants during first few months of life
- apneic-like pauses of 5 - 10 secs followed by a burst 50 - 60 resp/minute for 10 - 15 secs
- *not associated with heart rate or color change*
- infants should have respiratory rate recorded for 30 second intervals x 2 to avoid falsely high or low counts

NORMAL CARDIAC FUNCTION

- normal heart rate in a neonate ranges from 80 - 180
- varying degrees of acrocyanosis is common
- newborns have a relatively thick right ventricular wall and elevated pulmonary vascular resistance (PVR)
  - right axis deviation (RAD) of the QRS on EKG is normal and can range from +125 to +180 degrees
  - QRS and T waves show small voltages
  - RV dominance results in tall R waves in V1, V2, and rV4
  - this PVR gradually decreases by 6 - 8 weeks and resembles that of an adult
- normal PMI is at the left lower sternal border
- early systolic murmur caused by a normal persistent patency of the ductus arteriosus may be heard in the first few days of life
- innocent pulmonary flow murmur can be heard radiating to sides and back, usually < or = II/VI
- peripheral pulses should be palpable in all normal neonates, including pulses in the feet
- normal cardiothoracic ratio (CT ratio) on CXR is greater than 0.50 (inspiration and thymus affect CT ratio)

NORMAL ABDOMINAL ANATOMY

- liver is palpable 1 - 2 cm below the right costal margin
- spleen tip may be palpable

NORMAL FEEDING PATTERNS

- *FORMULA FED INFANTS* - will consume 3 - 4 ounces every 3 - 4 hours by the end of the first week of life
- *BREAST FED INFANTS* - will generally empty a breast within 7 - 8 minutes, so it's preferable to switch the infant to the other breast at that point and let him finish nursing on the side that still contains a supply of milk
- some infants will stop middle-of-the-night feeds by 3 - 6 weeks old, while others continue until 4 - 8 mo old
- infants may want to continue to suckle after feeding, try a pacifier !

MAKE SURE CHILD IS ACHIEVING ADEQUATE WEIGHT GAIN !!

NORMAL STOOLING PATTERNS

- first meconium stool should be passed within 24 hours of birth; if stool is not passed, possible Herschsprung's or hypothyroidism
- after milk feedings start, transitional stools start on the 3rd - 4th day and are greenish-brown with milk curds
- typical milk stools follow after an interval of 3 - 4 days
  - breast fed stools will be stringy, loose, yellow and sweet smelling
  - formula fed stools will be pasty, homogenous, yellow-brown; can be foul smelling
- frequency of stools closely relate to the frequency and amount of feeds, usually 3 - 5/day
  - breast fed infants may stool after every feeding
- No two infants are alike!!!

NORMAL GENITOURINARY ANATOMY

- infants born in the breech position may have significant bruising to the genital area

NORMAL VITAL SIGNS

AGE	HR (2%) mean (98%) mean +/- 2SD	BP Systolic	BP Diastolic	RR
Newborn		60 +/- 1**X**	37 +/- 8	40
1 - 2 days	(91) 123 (159)			< 40
3 - 6 days	(91) 129 (166)			
1 - 3 weeks	(107) 148 (182)			
1 - 2 mo	(121) 149 (179)	80+/- 16	46 +/- 16	24 - 35

NORMAL HEMATOLOLOGY VALUES

AGE	HGBgm% mean (-2SD)	HCT%	MCV	RETIC%	WBC
newborn	13.7 - 20.1				
1 - 3 days	18.5 (14.5)	56 (45)	108 (95)	1.8 - 4.6	18.9 (9.4 - 34)
2 weeks	16.6 (13.4)	53 (41)	105 (88)		11.4 (5 - 20)
1 mo	13.9 (10.7)	44 (33)	101 (91)	0.1 - 1.7	10.8 (4 - 19.5)
2 mo (nadir!)	11.2 (9.4)	35 (28)	95 (84)		
6 mo	12.6 (11.1)	36 (31)	76 (68)	0.7 - 2.3	11.9 (6 - 17.5)

- Normal term newborn hemoglobin = 13.7 - 20.1 gm/dl
- Physiologic nadir occurs at 8 - 12 weeks old; hemoglobin = 11.4 +/- 0.9 gm/dl; then erythropoiesis resumes
- Percentage that is fetal varies from infant to infant; fetal hemoglobin gone by 7 months old

NORMAL CSF VALUES

CELL COUNT	term 0 - 28 days	0 - 22WBC/mm3 (mean 8.2)	predominant lymphs
	> 1mo old	0 - 7 WBC/mm3	0% PMN
GLUCOSE	term infant	34 - 119mg/dl (mean 52)	
	child	40 - 80mg/dl	
PROTEIN	term infant	20 - 170mg/dl (mean 90)	
	child (lumbar)	5 - 40mg/dl	

- VS, Hematology, CSF values modified from The Harriet Lane Handbook 13th Edition

NEWBORN SCREENS and TREATMENT:

AAP GUIDELINES STATE ANY NEWBORN DISCHARGED PRIOR TO 48 HOURS OLD BE RE-EVALUATED WITHIN 2 - 3 DAYS !!!

- Phenylketonuria (PKU)
- Congenital Hypothyroidism
- Beta Thalassemia
- Galactosemia
- +/- other inborn errors, sickle cell, congenital toxoplasmosis, congenital adrenal hyperplasia, cystic fibrosis
- Coomb's test for infants of Rh (-) and "O" mothers
- Vitamin K 1mg IM
- Erythromycin ointment, Silver nitrate, (or diluted Betadine) to the eyes

NEONATAL RESUSCITATION EQUIPMENT:

- ETT TUBE 2.5, 3.0, 3.5, 4.0
- ETT BLADE 0 or 1 straight blade
- CHEST TUBE 10 - 12 F
- NG TUBE 6 - 8 F
- FOLEY TUBE 5.0 feeding tube, or 6 - 8 F Foley
- UMBILICAL VEIN CATHETER 3.5 or 5 F catheter or feeding tube
- CENTRAL LINE 4.0 F
- INTRAOSSEOUS with T-connector / 3 way stopcock
- MEDICATIONS

NIGHTMARE NEONATE

• term infant, with usually normal Apgars, who suddenly deteriorates in the first 1 - 2 weeks of life after a well interval at home ----> presents to the ED in extremis !!

COMMON CAUSES OF A CRASHING NEONATE

S = Sepsis
- Viral (Herpes, Enterovirus)
- Bacterial (E.coli, Strep, Listeria)

I = Inborn Errors of Metabolic Origin

C = Congenital cardiac disease
- PDA dependent left-outflow obstruction lesions
- Large left-to-right shunts
- Cardiomyopathies

C = Congenital Adrenal Hyperplasia (CAH)

C = CNS hemorrhages
- AV malformation
- Child abuse
- Vitamin K deficiency

F = Formulas mixups

I = Intestinal disasters
- Volvulus
- Necrotizing enterocolitis

T = Toxins and other home remedies

• modified, in part, from Michael Simmons MD - Harbor/UCLA

NEONATAL SEPSIS

• "Early onset" - seen in first few days of life, associated with maternal or perinatal risk factors, such as maternal fever, PROM, and fetal distress; septic shock and neutropenia more common presentation
• "Late onset" - usually occurs after 1 week of age, develops more gradually, less associated with above risk factors; meningitis more common presentation

CAUSES OF NEONATAL SEPSIS

- Group B streptococcus (most common in US)
- Listeria monocytogenes, E. coli, Klebsiella, enterococcus, nongroup D alpha hemolytic strep, and nontypable Haemophilus influenzae
- viral causes include herpes simplex, enterovirus (coxsackie, ECHO), and adenovirus (hepatic and CNS usually involved)

** Group B strep and Listeria can present early (<72 hrs) with sepsis, or late (4 - 14 days) with meningitis

CLINICAL SIGNS OF NEONATAL SEPSIS

"Not doing well"
Lethargy, irritability, seizures
Poor feeding, vomiting, diarrhea
Temperature instability (high or low)
Abdominal distension (ileus)
Apnea, tachypnea, cyanosis, respiratory distress
Hypoglycemia, hyperglycemia
Jaundice, pallor, petechiae
Tachycardia, bradycardia
Low blood pressure, poor perfusion
hepatosplenomegally
Congestive heart failure

ABDOMINAL DISTENSION MAY BE THE FIRST SIGN OF A SEPTIC NEONATE !!

ED WORKUP

- ask mom about maternal viral symptoms - check for asymptomatic herpes in mother
- Herpes Simplex -
  - usually presents as
  (1) disseminated infection involving multiple organs presenting as irritability, seizures, respiratory distress, jaundice, coagulopathy and shock
    - 20% will *not* have a vesicular rash
  (2) encephalitis with or without the skin lesions
    - CSF will show pleocytosis and elevated protein
  (3) disease localized to skin, eyes, or mouth
    - least frequent of all forms of disease
  - neonatal infection usually associated with primary herpes in the mother
  - examine neonate for herpetic lesions - usually occurs at the birth "presenting" portion of the body
  - check scalp electrode sites for herpetic lesions
- culture all body fluids, including CSF
  - less likely to make the child hypoxic if LP done in sitting position or lateral nonflexed
  - place infant on a pulse oximeter while LP is being performed
  - obtain blood glucose (prior to lumbar puncture) to compare with CSF glucose and to rule out hypoglycemia
- send urine culture even if dipstick, micro negative

- chest Xray only if symptomatic ??
- RSV and Pertussis in the first month of life can result in apnea so ADMIT to monitored bed !

LAB RESULTS

- Neutropenia (< 2,000 PMN/mm3), neutrophilia (> 16,000 PMN/mm3) or elevated immature-to-total neutrophil ratio (> 0.2) can be useful in predicting sepsis
- platelet count may be low in infants with sepsis
- CSF showing WBC's or high protein, but no organisms ---> think HERPES !

ED TREATMENT

☛ *ALL NEONATES WHO ARE WORKED UP FOR SEPSIS SHOULD BE ADMITTED !!*

- IV Ampicillin 100 - 200 mg/kg/24hr (Gram positives, Listeria, enterococcus) and Gentamicin 5 mg/kg/24hr (synergism plus broad gram negative coverage)
  OR Ampicillin and Cefotaxime (100mg/kg/day)
- If patient has positive CSF, use IV Cefotaxime for better CNS penetration
- Consider Acyclovir 10mg/kg per dose q 8 hours IV if WBC's or high protein but no organisms on CSF, pleocytosis, vesicular rash on infant, focal neurological signs, pneumonitis or hepatitis are present, or if there is a positive maternal history for herpes

CARDIOLOGY

A BRIEF INTRODUCTION TO CONGENITAL HEART DISEASE PRESENTING IN THE NEONATAL PERIOD !!

- congenital heart disease (CHD) occurs in 8/1000 live births

☛ *Most cardiac emergencies presenting in the neonatal period will present as cyanosis, cardiovascular collapse, congestive heart failure, or as an arrhythmia !!*

CLUES TO CONGENITAL HEART DISEASE

BLUE ---> cyanotic heart disease with *right to left* shunting
MOTTLED or GRAY ---> outflow obstruction with systemic hypoperfusion and shock
PINK ---> congestive heart failure with *left to right* shunting

AGE OF PRESENTATION

PDA-dependent lesions - cyanotic or shock-producing cardiac lesions- usually have sudden onset and usually present in first week of life
CHF lesions - usually have slower onset and present in late neonatal or early infancy period

• modified from Tintinalli 4th Edition

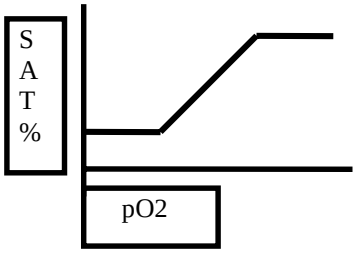
CYANOTIC HEART DISEASE:

Cyanosis implies 5 gm/dl of deoxygenated (reduced) blood or abnormal pigment like methemoglobin

- becomes deoxygenated due to decreased arterial saturation or increased extraction of oxygen by sluggish blood (shock, hypovolemia, or vasoconstriction)

0xygen-Hemoglobin Dissociation Curve -

- low pO2 leads to large drops in hgb saturation (affinity) so O2 can be delivered more effectively to the tissues
- shift to right (incr temp, incr pCO2, decr pH, incr 2,3 DPG) for a given pO2, better delivery to the tissues
- shift to left (fetal hgB)
- for a given pO2, have higher O2 sats and less delivery to tissues
- curve shifts to right like adult hemoglobin at approx 3 mo of age



Cyanosis is based on amount of deoxygentated blood and not the percentage,

- normally deoxygenated hgb = 2 g/dl in the venules; need another 3 g/dl to appear cyanotic
- polycythemic infants (neonates) may be cyanotic but still delivering O2 to the tissues
  ex: hgb 20 g/dl ---> if dexoxy 3 g/dl ---> oxygenated hgb 17/20 = 85% oxygenated
- anemic infants may not appear cyanotic yet still hypoxic, and not delivering O2 to the tissues
  ex: hgb 6 g/dl ---> deoxy 3 ---> oxygenated hgb 3/6 = 50% oxygenated
- may not appear cyanotic until the 02 sats drop to 50%

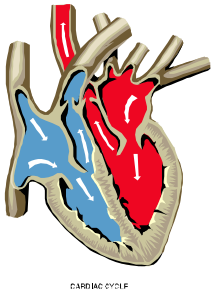
If hemoglobin and cardiac output are normal, a right-to-left shunt must be present to produce cyanosis; this can be either intracardiac, intrapulmonary or both !!

DIFFERENTIAL DIAGNOSIS FOR NEONATAL CYANOSIS

Cyanotic congenital heart disease
Parenchymal pulmonary disease
Diaphragmatic hernia
Persistant pulmonary hypertension of the newborn
Polycythemia
Hypoglycemia
Shock and sepsis
Central Nervous System Disease
Hemoglobinopathy
Congenital Methemoglobinemia

CONGENITAL HEART DISEASE causing CYANOSIS

Tetralogy of Fallot (TOF) - may be overlooked in nursery
Tricuspid atresia
Transposition of the Great Arteries (TGA)
Total Anomalous Pulmonary Venous Return (TAPVR)
Truncus Arteriosus - may be overlooked in the nursery
Pulmonary atresia or stenosis
other less common lesions



• modified from Donn Neonatal Emergencies

*** These congenital heart defects produce cyanosic (hypoxemia) because of right-to-left intracardiac shunting*
*** Many of these lesions may be dependent upon the ductus arteriosus to remain patent and maintain blood flow to the lungs ---> when the PDA finally closes, the child may suddenly become noticeably cyanotic !!*

CLINICAL SIGNS

• previously healthy neonate suddenly presents with lethargy, pallor or central cyanosis
• usually presents in the 1st week of life (up to 3 weeks old)
• history reveals difficulty feeding, poor weight gain, tachypnea, and diaphoresis
  - feeding times are the neonate's cardiac stress test !!
• differentiate central vs peripheral cyanosis -
  - peripheral cyanosis will still have pink tongue, mucous membranes (hard to evaluate lips)
  - can be difficult to evaluate in presence of acrocyanosis, darker skin, and artificial light
• neonate may appear quite comfortable with only mild tachypnea and cyanosis and no respiratory distress
  - infant is often described as "happily tachypneic"
  - cyanosis due to pulmonary pathology will usually present with retractions, nasal flaring, and grunting
• cardiac cyanosis will worsen with crying, improve with rest
  - pulmonary cyanosis will improve with crying due to increased ventilation

DON'T ASSUME THAT ACROCYANOSIS IS NORMAL IN A LETHARGIC NEONATE !!!

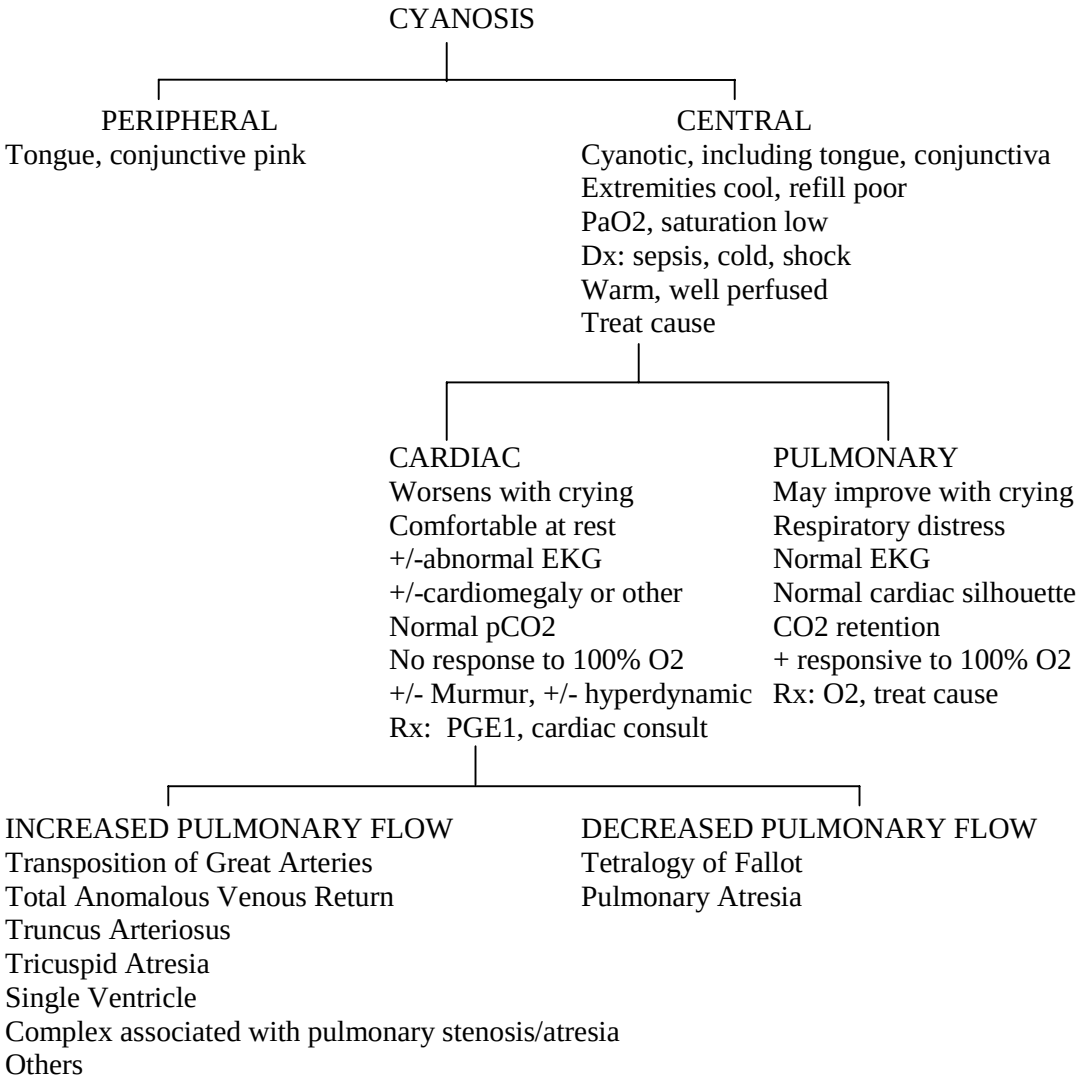
• cardiac exam - pulses typically normal and equal; precordium is quiet; may not have a murmur
  - pulmonary stenosis - right ventricular lift and thrill
  - Tetralogy of Fallot, truncus arteriosus - murmur usually

• ABG - will show decreased PaO_2
  - Hyperoxitest - 100% O_2 will not alter the ABG (allow at 10 min for O_2 effect)
  - a rise of > 30 torr or pO_2 > 100 - 150 is highly suggestive of lung disease, and not cardiac disease but must be interpreted in light of clinical situation
  - CO_2 retention suggests pulmonary or central nervous system disease
  - low pH suggests sepsis, shock or severe hypoxemia

• EKG - usually non-diagnostic because of normal neonatal RAD and dominant R wave in right chest leads
  - Tricuspid atresia will show LAD and LVH since right heart not well developed
  - T wave also upright in V1 for first 4 days; beyond 4 days consider pathology

• CHEST XRAY - should include heart size and position, liver shape and position, and increased or decreased vascularity
  - Cardiothoracic ratio can be hard to evaluate due to thymus size and depth of inspiration
  - CT ratio usually greater than 0.5 in normal neonate without CHD
  - cardiomegally due to cong heart disease may not manifest yet in the newborn period
  - if unequivocal cardiomegally, common causes include:
    - cong heart disease - VSD, PDA, TGA, Hypoplastic left heart, Ebstein's anomaly
    - myocarditis, cardiomyopathy
    - pericardial effusion
    - metabolic disturbances like hypoglycemia or acidosis
    - overhydration or overtransfusion
  - abnormal cardiac silhouette-
    - "boot shaped" - TOF or tricuspid atresia
    - "egg shaped" - TGA
    - large globular heart - Ebstein's anomaly
  - dextrocardia or mesocardia can be a sign for congenital heart disease
  - location of stomach bubble, shape and location of liver can also be a marker for CHD
  - pulmonary vascular markings -
    - cyanotic with *decreased* vessels - TOF, Pulmonary atresia, Tricuspid atresia
    - cyanotic with *increased* vessels - TGA, Truncus arteriosus, Single ventricle, TAPVR
    - *acyanotic* with increased vessels - VSD, PDA, endocardial cushion defect

ALGORITHM FOR EVALUATION OF CYANOTIC NEONATE (Flynn 1992)



ED TREATMENT

- AIRWAY, BREATHING, CIRCULATION !!
- Oxygen and ventilatory support as needed
  - even a small rise in p02 may be of benefit
- If ductal-dependent cardiac lesion is suspected,

  Prostaglandin E1 (PGE1) 0.05 - 0.1 mcg/kg/min IV
  - 500 mcg/100 ml NS = 5 mcg/ml ---> start infusing 0.1 mcg/kg/min = 0.02 ml/kg/min
  - potent vasodilator --> ductal tissue very sensitive to its action
  - O2 sats usually rise to 80 - 90%, sometimes up to 100%
  - neonate now at risk for apnea (12%), so consider intubation, especially if inter-hospital transport is necessary
  - other side effects include hypotension, seizures, fever, jitteriness
  - results usually within 15 minutes
  - can increase rate up to 0.4 mcg/kg/min if needed for effect
  - after cyanosis is relieved (or BP improved) , lower PGE1 rate down slowly by small increments to 0.01 mcg/kg/min
- Any neonate this ill deserves a septic workup and antibiotics until sure of diagnosis !!
- Admit to Neonatal or Pediatric ICU

• Echocardiogram
CONSULT A PEDIATRIC CARDIOLOGIST OR THE NEONATAL ICU IN YOUR AREA!! ☎

CARDIOVASCULAR COLLAPSE:

• usually occurs in the first 2 weeks of life; neonate had been discharged prior to ductus constriction, without obvious signs:
- perfusion and pulses were maintained by the PDA
- PDA was so wide, a murmur wasn't audible at birth

CONGENITAL HEART DISEASE causing CARDIOVASCULAR COLLAPSE
Coarctation of the Aorta or Interrupted Aortic Arch
Hypoplastic Left Heart
Critical Aortic Stenosis

☛ ** Patent ductus had allowed adequate right to left blood flow to the sytemic circulation prior to its closure ---> after PDA closure, greatly diminished sytemic blood flow results !*
* Can also result in congestive heart failure with pulmonary edema

CLINICAL SIGNS

• upon PDA closure, neonate presents in shock with a history of poor feeding, tachypnea and poor color
• pale, clammy, hypotension, diminished/absent pulses, mottling, poor perfusion, lethargy
• aortic stenosis, hypoplastic left heart ---> poor pulses throughout
- aortic stenosis ---> thrill, murmur
- hypoplastic left heart - cardiomegaly
• coarctation - ductus usually positioned right at the area of coarctation and adds to aortic lumen size
- closure of ductus allows even less blood flow through the aortic coarctation
- absent or delay in pulses to the lower extremities is most common presentation
- rarely, upper body may be pink (precoarc) and lower body may be blue (postcoarc)
- check blood pressures/O2 sats/pulses in the right arm and leg
- a difference in p02 of 10 - 15 mmHg is significant
- left subclavian artery can be variably pre or post-ductal so don't use left arm
- difference in peripheral blood pressures may not be apparent until after inotropes administered
- may have an associated VSD which may cause mixing of oxygenated and deoxygenated blood
• interrupted aortic arch ---> incr pulses in right upper extremity; poor pulses in the left upper and both lower extremities
• pulmonary exam usually positive for dyspnea and pulmonary congestion (usually wheezing, not true rales)
• CXR will usually show cardiomegaly and pulmonary edema
- coarctation may have a "3" sign on plain Xray or "E" on barium swallow
• EKG typically shows RVH or RBBB (older infants will show LBBB)

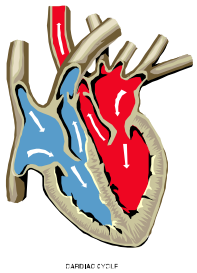
ED TREATMENT

• see treatment for "Cyanosis"
• anticongestive agents like inotropic agents (dopamine, dobutamine) and diuretics should be started

CONGESTIVE HEART FAILURE:

DIFFERENTIAL DIAGNOSIS FOR NEONATAL HEART FAILURE

Congenital heart disease
Myocarditis
Arrhythmia (SVT or complete heart block)
Arteriovenous fistula (intracerebral, intrahepatic)
Asphyxia
Hypoglycemia; Hypocalcemia
Anemia
Sepsis



CONGENITAL HEART DISEASE causing CONGESTIVE HEART FAILURE

Large left-to-right intracardiac shunt - excessive pulmonary circulation (more common)
- Ventricular septal defect, large
- Arterio-venous malformations
- Complete AV canal
- Patent ductus arteriosus, large

Diminished left ventricular function (less common)
- Myocarditis
- Dilated cardiomyopathy
- Anomalous origin of left coronary artery from the pulmonary artery

• modified from Donn Neonatal Emergencies

** Normal transitional circulation after birth includes a gradual decline in pulmonary vascular resistance --->this allows a pre-existing left-to-right shunt to progressively increase its flow*
• neonate usually presents after 2 weeks of age, as late as several months
• symptoms present more gradually, often insidiously; murmur may not present until 2 weeks - 2months old

CLINICAL SIGNS

• *CHF signs are due to pulmonary overcirculation and sympathetic stimulation, or due to left ventricular failure !*
• poor feeding, diaphoresis (especially while eating), tachypnea, tachycardia, hepatomegaly, cardiomegaly and finally, pallor, mottling, hypotension, and oliguria
• *difficulty feeding is a fairly constant feature, with the neonate tiring and diaphoretic*
• heart rates above 180 - 200 when the neonate is at rest suggest increased autonomic activity to compensate for a failing myocardium
• respiratory rates above 50 - 60/ minute, usually without increased depth, is an early sign
• grunting, flaring of the nose, and retractions are unusual unless there is pulmonary disease or frank pulmonary edema
  - often an intercurrent illness is what tips the infant over the edge
• neck veins are usually not discernible in a neonate
  - other peripheral edema signs and ascites are unusual in the neonate, sometimes sacral edema present
  - "hepatomegaly" usually due to depressed diaphragm due to pulmonary hyperinflation
• wheezing may be heard; rales are unusual
• cardiac exam - usually has an hyperactive precordium
  - significant left-to-right shunts ---> diastolic flow rumble at the apex

- large PDA - classic continuous “machinery” murmur, with wide pulse pressure
• AV malformation may be heard over anterior fontanelle or liver
• CXR usually shows cardiomegaly, increased vessels, and fluid in the minor fissure

ED TREATMENT

• AIRWAY, BREATHING, CIRCULATION !!
• Oxygen and ventilatory support as needed
• Lasix 1mg/kg IV
• Dopamine or Dobutamine for systemic hypotension
• Echocardiogram
• Any neonate this ill deserves a septic workup and antibiotics until sure of diagnosis !!
• Admit child to a Neonatal or Pediatric ICU

CONSULT A PEDIATRIC CARDIOLOGIST OR THE NEONATAL ICU IN THE AREA !! ☎

INBORN ERRORS OF METABOLISM

PATHOPHYSIOLOGY

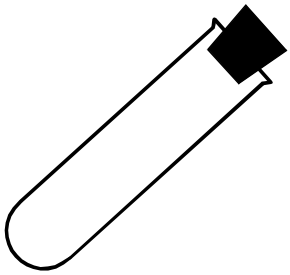
• Inborn errors refer to the hundreds of hereditary biochemical disorders resulting in the alteration of a protein structure or amount being synthesized
• usually the result is a deficiency of an enzyme needed in the conversion of one metabolite to another ---> results in the accumulation in body fluids of a metabolic intermediate that normally is present in low concentration
  - normally these metabolic intermediates are not toxic but high levels can cause serious effects
  - usual target organ affected is the central nervous system
• some of these biochemical disorders are clinically inconsequential, and others range from mild to lethal
• most inborn errors manifest themselves in the newborn period or soon after

CLINICAL SIGNS

• lethargy, coma, failure to thrive, or just persistent vomiting as the only symptom !
• seizures
• hepatomegaly with or without icterus or coagulopathy
• metabolic acidosis with or without ketosis or ketonuria, often with hyperpnea
  - acidosis present only in the organic acidemias, unless other causes exist like dehydration, or cardiac arrest
• elevated blood or urine levels of a particular metabolite, like an amino acid or ammonia
• a peculiar odor
• sepsis with or without bone marrow suppression
• intraventricular or pulmonary hemorrhages are frequent agonal events
  - look for signs of increased intracranial pressure
• ask about previous siblings, especially males, who died in infancy !!

☛ *A HISTORY OF CLINICAL DETERIORATION IN A PREVIOUSLY NORMAL NEONATE SHOULD SUGGEST SEPSIS AND /OR AN INBORN ERROR OF METABOLISM !!*

ALGORITHM FOR INBORN ERRORS OF METABOLISM



Initial findings include:
- poor feeding
- vomiting (may be the only symptom !!)
- lethargy
- convulsion- not responsive to IV glucose or calcium
- coma- not responsive to IV glucose or calcium

METABOLIC DISORDER — INFECTION

Obtain plasma AMMONIA

High — Normal

Obtain blood pH, CO2 — Obtain blood pH, CO2

Normal — Acidosis — Normal

UREA CYCLE DEFECT — ORGANIC ACIDEMIAS — AMINOACIDOPATHIES OR GALACTOSEMIA

• Nelson's TEXTBOOK OF PEDIATRICS 14th Edition

ED TREATMENT

- Admit to an observation unit or ICU
- goal is to limit production of more toxic metabolite and encourage elimination via hydration and adequate glucose
- rehydrate the infant with adequate boluses of normal saline 20cc/kg IV
- maintenance fluid then at one and a half to two times normal maintenance rate
- follow glucoses and bolus as needed with 0.5 - 1g/kg (D 25 2 - 4cc/kg) IV
- D10 as constant infusion will stimulate insulin production and protein synthesis
- Labs include electrolytes, glucose, calcium, ketones, ABG, urinalysis
- ammonia level (best if arterial, green top/heparin tube, on ice)
- lactic acid if metabolic acidosis is found (7 ml green top, on ice)
- urine may show spindle shape orotic acid crystals if urea cycle defect exists
- save urine and plasma in freezer for further studies (red and green top blood tubes)
- treat seizures with benzodiazepines
- consider HCO3 for pH < 7.1 (but alkalinization can increase ammonia passage into CNS)
- any neonate presenting this ill should also have a sepsis workup and appropriate antibiotics
- contact the NICU/PICU in your area to discuss "antidote therapy" like sodium benzoate for ammonia and transport

CONGENITAL ADRENAL HYPERPLASIA

- occurs in 1/10,000 - 1/15,000 live births; much higher in Alaskan Yupik Eskimos (1/300)

PATHOPHYSIOLOGY

- Adrenal insufficiency resulting from deficient activity of one of the five enzymes required to produce cortisol
- most common enzyme deficient is 21-hydroxylase enzyme

- results in decreased conversion of 17-OH progesterone ---> 11-desoxycortisol in the glucocorticoid pathway
  - this leads to a *deficiency in cortisol* synthesis, and subsequent hyperplasia of the adrenal gland as a result of overstimulation by ACTH (has no negative feedback by cortisol)
  - cortisol deficiency results in cardiovascular collapse

- most also have decreased conversion of progesterone ---> 11-desoxycorticosterone in the mineralcorticoid pathway
  -this leads to a *deficiency of aldosterone* synthesis, leading to urinary salt wasting
  - aldosterone deficiency results in classical electrolyte findings and contributes to cardiovascular collapse

- as a result of elevated ACTH stimulation, adrenal *steroid precursors accumulate* and are metabolized to *androgens* ---> resulting in the virilization of the external genitalia in female infants
  - since male infants genitalia usually are not affected, may go unrecognized at birth !!

CLINICAL SIGNS

- total body salt depletion, vomiting, dehydration that may lead to circulatory collapse and death during the initial 2 - 3 weeks of life (commonly at the end of the first week of life)
- females may have enlarged clitoris and fusion of labial folds; males may have small phallus

LAB RESULTS

- hyponatremia, hyperkalemia, azotemia, and metabolic acidosis
- can also cause hypoglycemia

ED TREATMENT

- Admit to an PICU or NICU
- labs - 17 hydroxyprogesterone, dehydroepiandrosterone, androstenedione, testosterone if possible before giving hydrocortisone (red top tube, 5 - 6 ml total)
- volume repletion with 0.9% normal saline (20cc/kg boluses) then D5 0.9% NS at 100 - 125 ml/kg/24hr
- cortisol replacement with hydrocortisone 25 mg IV bolus, then 25 - 50 mg/m2/day divided q 6 - 8 hrs
- extreme hyperkalemia usually well tolerated and saline is usually only measure needed to lower K+, but may use IV 10% calcium gluconate for arrhythmias
- Remember to monitor temperature and glucose also !!

CNS DISASTERS and SEIZURES

NEONATAL SEIZURES

- Generalized tonic/clonic, jacksonian march and absence seizures are rarely seen in neonates
- Electrical discharges are incompletely spread and tend to remain localized due to anatomic and physiologic CNS immaturity
- Many neonatal seizures involve subtle motor automatisms (see below)
- "Electroclinical dissociation" is common - clinical seizure but no EEG correlation
- Few idiopathic seizures in neonates*; SEARCH FOR ETIOLOGY IS MANDATORY !*
- Focal seizures can be caused by metabolic disorders, and do not necessarily imply a focal CNS lesion
- Evidence to suggest that the seizures themselves may be damaging to the developing neonatal brain
- Occurs in 0.2 - 1.4% of all newborns; mortality ranges from 15 - 40%

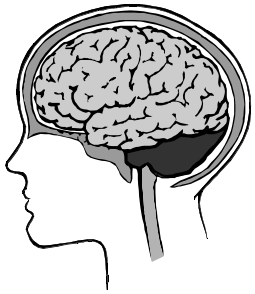
DIFFERENTIAL DIAGNOSIS OF NEONATAL SEIZURES

Central Nervous System

Hemorrhage - subdural, intracortical, intraventricular (15 - 20%) *
- rule out hemorrhagic disease, ABUSE !!
Hypoxic encephalopathy/ birth trauma (30 - 65% of cases) *
Congenital anomalies or developmental brain disorder *
Cerebral necrosis/ infarcts
Cortical vein thrombosis

Metabolic and Systemic

Hypertension
Hypocalcemia *
Hypoglycemia *
Hypomagnesemia
Electrolyte imbalance (hyper or hyponatremia) *
Inborn Errors of Metabolism *
Hyperthermia
Pyridoxine (B6) deficiency/dependency *
- maternal use of INH



Infections (10 - 15%)

Bacterial Meningitis *
Cerebral Abscess
Herpes Encephalitis *
Coxsackie meningoencephalitis
Congenital (Cytomegalovirus, Toxoplasmosis, Syphilis) *

Drug Withdrawal *

Methadone
Heroin
Barbiturates
Propoxyphene

* = unique or of concern in neonatal period

Toxins

Local anesthetic
Bilirubin *

Familial seizures

Benign familial neonatal seizures *
Benign idiopathic neonatal ("5th day fits") seizures *

CLINICAL SIGNS

- seizures may be subtle in the neonate i.e. staring spells, prolonged eye deviation, nystagmus, lip smacking, tongue thrusting, bicycling, brief altered muscle tone, apnea, or autonomic changes (BP fluctuation, tachycardia, pupil dilation)
- clonic seizure may be focal and migratory (first one leg, then opposite arm); consciousness usually maintained
  - clonic movement is slower and more rhythmic than an older child's clonic movements
- tonic seizure with hyperextension of trunk, neck or limbs is another variant
- convulsive apnea, unlike non-convulsive paroxysms, thought not associated with bradycardia (Fenichel 1983; although Watanabe 1982 did not confirm this)
- neonatal jitteriness (non-seizure) will involve fast movements of all extremities, stimulation will induce movements, movements will stop with restraint or passive flexion, eye movement and gaze are normal, and rarely has autonomic signs or symptoms

CLUES TO NEONATAL SEIZURES BASED ON AGE AT PRESENTATION:

First 48 hours - trauma, pyridoxine dependency, hypoxic encephalopathy, hypoglycemia
- benign familial neonatal seizures - usually begin on 2nd - 3rd day, resolve by 1 - 6 months

4 - 7 days old - hypocalcemia due to high phosphate load from formulas
- benign idiopathic "fifth day fits" - start on 5th day and cease by day 15

> 7 days old - infection

ED WORKUP

- History - prenatal and labor history regarding infection risks, prenatal TORCH studies, substance abuse, perinatal asphyxia, or family history of seizures
  - opiate withdrawal seizures can present up to several weeks post-birth
  - if bottle fed, how does mom mix formula; has mom supplemented or replaced infants diet with free water or tea or other home remedies ex baking soda for colic
- Physical Exam - blood pressure check for hypertension
  - unusual odor of sweat or urine for inborn errors
  - cranial bruits for AVMs
  - skin for jaundice, cafe-au-lait spots, herpes vesicles (look at scalp electrode sites)
  - neuro exam for cranial nerves, motor exam, neonatal reflexes
- Potential labs - glucose, hematocrit, electrolytes, BUN, calcium, magnesium, phosphate, serum ammonia, ABG, blood cultures (bact and viral), TORCH titers
  (send calcium and magnesium even if seizures ceased by anticonvulsants)
  - urine for urinalysis and toxicology screen
  - urine for 2,4-dinitrophenilhydrazine and reducing substances
  - blood for amino and organic acids, lactate and pyruvate (green top tube in freezer)
  - urine for amino and organic acids, lactate and pyruvate (save in freezer)
  - CSF for glucose, protein, cell count, diff, gram stain, bacterial and viral cultures, latex agglutination for viral antigens, lactate and pyruvate, glycine
- Ultrasound of cranium plus CT or MRI scan

TREATMENT

- AIRWAY, BREATHING, CIRCULATION !
- bedside glucose check
  - administer a glucose bolus 0.5 - 1.0 grams/kg (2 - 4cc/kg) D25 IV if indicated
- standard anticonvulsant therapy such as benzodiazepines, phenobarbital or dilantin indicated
  - most NICUs will recommend benzodiazepines or phenobarbital as first line drug
  - Phenobarbital 18 - 20 mg/kg IV (infuse no faster than 1 mg/kg/min); may repeat 5 mg/kg/dose q 5 - 10 minutes, up to total dose of 40 - 60 mg/kg
  - Lorazepam (Ativan) 0.1 mg/kg IV or rectally over 2 minutes, may repeat 0.05 mg/kg IV x one
  - Diazepam (Valium) 0.5 mg/kg IV or rectally q 15 - 30 min x 2 - 3 doses
  - ? sodium benzoate theoretically can displace bilirubin from albumin --> at risk for kernicterus
  - Dilantin 15 - 20 mg/kg IV (infuse no faster than 1mg/kg/min)
  - ?? Paraldehyde 0.3ml/kg diluted 1:2 in mineral oil given rectally
- consider infusing calcium gluconate 10% 100 - 300 mg/kg IV (1 - 3 ml/kg at 1 ml/min) if still seizing after standard therapy, or has a weak cry (like a bleating lamb)
- consider infusing magnesium sulfate 50% 0.1 - 0.3 ml/kg IV or IM
- consider infusing pyridoxine (B6) 50 - 100 mg IV if still seizing after standard anticonvulsant therapy, glucose, and calcium infusion
  - pyridoxine is a cofactor for the synthesis of inhibitory neurotransmitter GABA
  - need EEG monitoring; will see clinical response within minutes
- treat hypertension induced seizures with antihypertensive meds
- antibiotics (i.e. Cefotaxime and Ampicillin) to the septic patient
- consider Acyclovir 10mg/kg per dose q 8 hours IV if WBC's or high protein but no organisms on CSF, pleocytosis, vesicular rash on infant, focal neurological signs, pneumonitis or hepatitis present, or maternal history of herpes
- ADMIT TO A MONITORED BED !!!

SUBSTANCE WITHDRAWL:

- most commonly described with heroin, methadone, and morphine
- can also be seen with demerol, codeine, propoxyphene, and pentazocin even if not chronically abused
- onset of symptoms is usually between 24 - 48 hrs old, but as late as 2 weeks if newborn is exposed to methadone; up to 34 days for heroine (classically day 10)
- classical symptoms include excessive irritability, decreased sleep time (can be wrongly diagnosed as colic), fever, vomiting, diarrhea, SEIZURES !!

SIGNS and SYMPTOMS OF NEONATAL DRUG WITHDRAWL

W = wakefulness
I = irritability
T = tremulousness, temperature variation, and tachypnea
H = hyperactivity, high-pitched persistent cry, hyperacusis, hyperreflexia, hypertonus
D = diarrhea, diaphoresis, disorganized suck
R = rub marks, respiratory distress, rhinorrhea
A = apneic attacks, autonomic dysfunction, alkalosis
W = weight loss or failure to gain weight
L = lacrimation (AAP Committee on Drugs 1983)

ED TREATMENT:
- ADMIT TO A MONITORED BED
- if history is unreliable, consider sepsis, hypoglycemia, and hypocalcemia
- increase infants comfort (swaddling, pacifier, decrease environmental stimuli)
- phenobarbital used as needed

FORMULA MIXUPS and TOXINS

FORMULAS

☛ *IF YOU DON'T ASK HOW THEY ARE MIXING UP THE FORMULA, PARENTS WON'T ALWAYS TELL YOU!!*

- Formula varieties:
  - Ready-made - don't add water
  - Concentrated liquid - add 1 scoop powder for 1 ounce water
  - Powder forms - add 1scoop powder for 2 ounces water

TOXINS and other home remedies

☛ *IF YOU DON'T ASK ABOUT ADDITIONAL LIQUIDS, POWDERS, HERBS, or OTHER SUBSTANCES GIVEN TO THE NEONATE, PARENTS WON'T ALWAYS TELL YOU !!*

- examples: baking soda for colic, herbal teas for constipation or colic

INTESTINAL DISASTERS

OMPHALITIS

• inflammation and infection can result in hematogenous spread or extension to the liver or peritoneum

CLINICAL SIGNS

• can have minimal signs such as mild erythema on abdomen, usually circumferential around umbilicus
• *ANY ERYTHEMA EXTENDING ONTO ABDOMEN IS OMPHILITIS UNTIL PROVEN OTHERWISE!!*

ED TREATMENT:

• Treatment - MEDICAL EMERGENCY !!!
• IV Oxacillin and Gentamicin and admission to observation area
• surgery necessary for abscesses

VOLVULUS

• Congenital malrotation of the midgut portion of the intestine - during the 5 - 8th week in embryonic life, the intestine projects out of the abdominal cavity, rotates 270 degrees and returns into the abdomen; if the rotation is not right, the intestine will not be "fixed down" correctly at the mesentery ------> at risk for malrotation
• Volvulus is the twisting of a loop of bowel about its mesenteric base attachment
• True medical emergency because necrotic bowel can occur within hours of onset of the twisting

CLINICAL SIGNS

• Generally peak occurrence in the first month of life but can present anytime in childhood; male to female 2:1 ratio; rarely familial
• Presents one of three ways:
  - sudden onset of bilious vomiting and abdominal pain
  - history of "feeding problems" with bilious vomiting that now appears like a bowel obstruction
  - failure to thrive with severe feeding intolerance (least common)
• BILIOUS (green) VOMITING IN NEONATES IS ALWAYS WORRISOME AND IS A TRUE EMERGENCY !!
• if bowel is already ischemic or necrotic, neonate may present pale and grunting
• abdomen may or may not be distended depending upon location of the volvulus; if obstruction is high, abdomen may not be distended; abdomen may be "blue" if bowel is already ischemic / necrotic
• pain is a constant pain, not intermittent
• neonate may be jaundiced
• hematochezia is a late, BAD sign !
• neonates present ill !!
• Differential:
  - Gastroenteritis - ill contacts ??; volvulus can appear like AGE early on; CAUTION
  - Pyloric stenosis - longer history; child acts well and hungry

ED WORKUP

• labs - nothing classic except dehydration and acidosis
• Abdominal plain film
  - classic "double bubble sign" - paucity of gas (airless abdomen) with two air bubbles - one in the stomach and one in the duodenum
  - plain film can also be entirely normal
• Upper GI - considered the gold standard
  - small intestine is rotated to right side of the abdomen; contrast narrows at site of obstruction "cork-screwing"; spiraling of small bowel about the superior mesenteric artery

- Ultrasound
  - may show a distended fluid-filled duodenum, increased peritoneal fluid and dilated small bowels loops to the right of the spine
  - Radiology 1996 - Shmianuki - Japan - Clockwise whirlpool sign of color Doppler - Japan - 236 children with suspicion for volvulus (day 0 to 14yo); whirlpool sign = wrapping of sup mes vein and mesentary around the sup mes artery; was clockwise in 12 / 13 kids with surgically confirmed volvulus; was counterclockwise in 3 without volvulus; sensitivity 92%, specificity 100%, PPV 100%
  - American Journal of Roentgenology October 1992 - 337 infants had utz for r/o HPS. Normally sup mes vein should be on right side of artery on transverse utz; in 74%, the anatomy could be seen; Nine were abnormal - 5 had vein on left side and all had malrotation; 4 had vein ventral to the artery and one had malrotation

ED TREATMENT

- Need to diagnose this life threatening process EMERGENTLY !!!
- Re-hydrate the infant aggressively; place an NG tube
- Antibiotics: Ampicillin, Clindamycin and Gentamicin
- When the diagnosis is being considered, contact the Pediatric Surgeon on-call immediately; the sooner the child gets to the OR, the lower the morbidity and mortality
  - some peds surgeons will take an ill appearing neonate with BILIOUS vomiting to the OR directly without any additional diagnostic tests
  - Journal of Formosan Medical Association April 1995 - Taiwan - 15 year retro review - bilious vomiting and bloody stools were more common in neonatal period; recurrent abdominal pain and FTT more common after newborn period; obscure symptoms and longer duration of symptoms were more common in the older child, leading to delayed diagnosis

SAFETY

• As recommended by the American Academy of Pediatrics infants should be placed on their sides or back while sleeping to decrease the risk of SIDS "BACK TO SLEEP!!"
  - smoking in the house increases risk of SIDS

• Infant should sleep in a regulated infant crib
  - slats should be no greater than "two adult sized fingers" apart
  - infant should not sleep on an adult bed - risk of falling off the bed, between the slats and asphyxiation, or risk being smothered by the mother
  - no pillows or plastics in the infants crib
  - infant should never be placed on a bean bag, water bed, sheep skin, or other soft beds

• Infant should never be left alone on a changing table or bed

• Infant should never, never, never be left alone in a bathtub; have a portable phone, install a phone in the bathroom or just let the phone ring

• When infant becomes mobile, never leave buckets of water unattended or toilets open

• Never leave a baby alone with a pet, even a well-behaved one

• Never leave a baby alone in a room with a sibling who is < 5 years old
  - a game of peekaboo could turn into tragic suffocation
  - an enthusiastic bearhug could break a rib

• Never leave the infant home alone
  - "I just stepped out for a second" can lead to tragedy

• In the car, infant must be placed in an appropriate car seat, and not held on parent's lap

• Never leave a child in a car alone
  - cars can overheat or someone can steal the infant

• Never take eyes off the child when in public, and be cautious when strangers offer to hold the child

• Never jiggle or shake the baby vigorously or throw him up in the air

• Infants less than 12 months old should not be allowed to eat grapes (fruits with skin), hard vegetables, hot dogs, popcorn, raisins, nuts, hard candies, lollipops, peanut butter, hard crusty bread, or hard meat
  - foods should easily dissolve if caught in the airway
  - children should be taught not to play or run while eating

• Infants should not be allowed to "bite" an inflated balloon ----> when the balloon explodes, the balloon pieces fly backwards into the infant's airway and occludes it

• Avoid using any kind of chain or string on the baby or on any of the toys or belongings
  - no necklaces, no chains, no string on pacifiers, no ribbon longer than five inches near crib

• Parent should know how to use and read a thermometer and be instructed to bring a neonate to the doctor for a temp > 100.4 degrees

• Modified from "WHAT TO EXPECT THE FIRST YEAR"

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