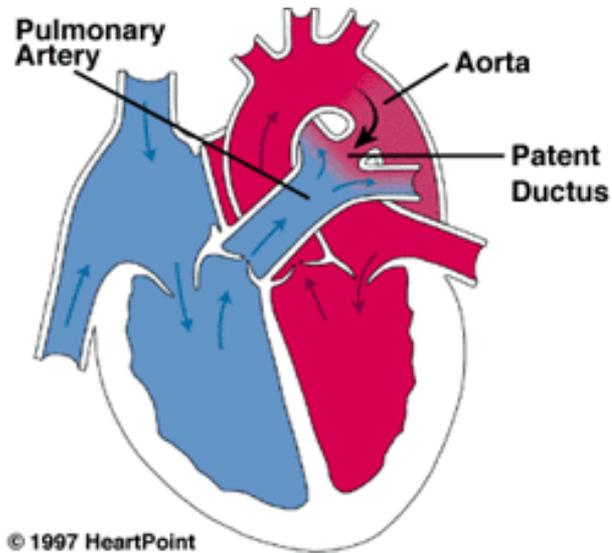


# Ductal-dependent cardiac anomalies



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Pediatric Emergency Rotation

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# Case 1

A 6 day old Caucasian male presents to the ER with poor feeding and SOB. He was born FT via VD with an uncomplicated antenatal period. APGARs were 9 and 9 for 1 and 5 min, BW was 3.2kg. He was sent home on day 2 of life for routine WCC in 1 week. He was well appearing but his skin was slightly “dark”/“had a black-purple hue” since birth. On discharge he initially took 2oz of Enfamil formula Q3H but gradually since then he has progressed to taking 1 oz of formula Q3H which takes him an hour to feed associated with SOB and occasionally sweating. He was also noted to be sweating excessively sometimes when he sleeps and has been sleeping a lot; when he is not trying to take the hour to feed, he is sleeping. When awoken he is very irritable.

# Case 1

His vitals on presentation are: T37.2C RR80 P170 BPs - RUL 68/40 LUL 65/39 RLL 67/41 LLL 68/39. Weight 2.7kg. O<sub>2</sub> at of RUL is 90% but of LLL is 58%. Examination does not reveal a murmur but he has a gallop rhythm, his liver is 2cm below the costal margin and all his pulses are equally reduced.

Of the following the MOST likely diagnosis is:

- A. Coarctation of the Aorta
- B. Transposition of the Great Arteries
- C. Hypoplastic Left Heart Syndrome
- D. Hypertrophic Cardiomyopathy secondary to Gestational Diabetes



# Ductal-dependent cardiac anomalies

- Neonates with congenital heart lesions may not be symptomatic during the birth hospitalization because the ductus arteriosus has not yet closed but they often present by two weeks of age.
- The lesions not diagnosed prior to discharge were primarily ductal dependent: coarctation of the aorta, interrupted aortic arch, aortic stenosis, hypoplastic left heart syndrome, and transposition of the great arteries.
- Closure of a patent ductus arteriosus can precipitate rapid clinical deterioration with potentially life-threatening consequences (ie, severe metabolic acidosis, seizures, cardiogenic shock, cardiac arrest, or end-organ injury).

# Ductal-dependent cardiac anomalies

Parents most commonly notice difficulty with feeding: intake of a limited volume of milk, feedings that are taking too long or are frequently interrupted by sleeping or resting, choking, gagging, +/- vomiting. Infants may have respiratory distress worse with feedings, or a persistent cough or wheeze.

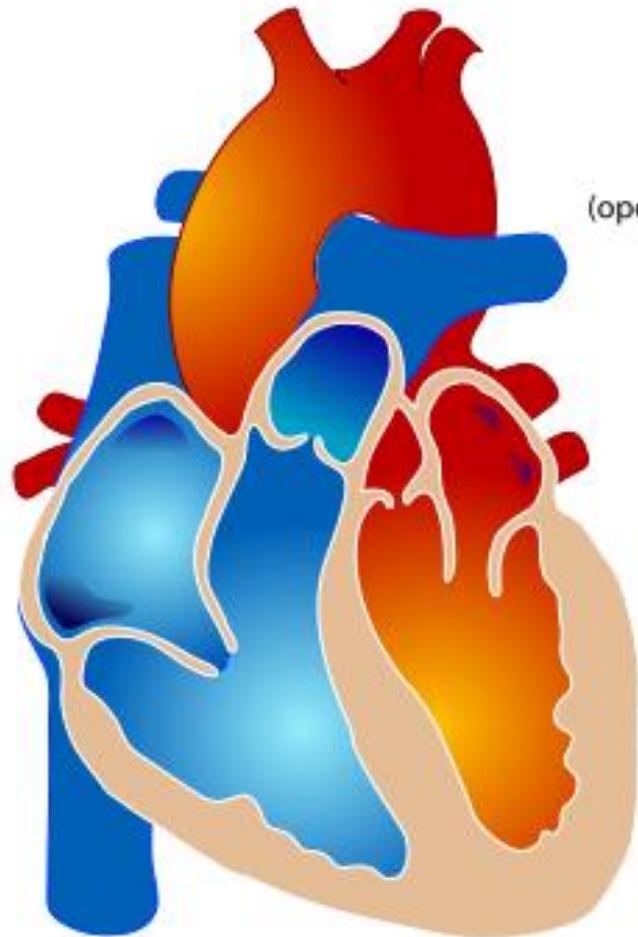
Other manifestations include:

- Color changes, such as central cyanosis or persistent pallor
- Excessive, unexplained irritability
- Excessive sweating that is increased with feeding and may occur during sleep
- Poor weight gain
- Decreased activity or excessive sleeping
- Delay in motor milestones

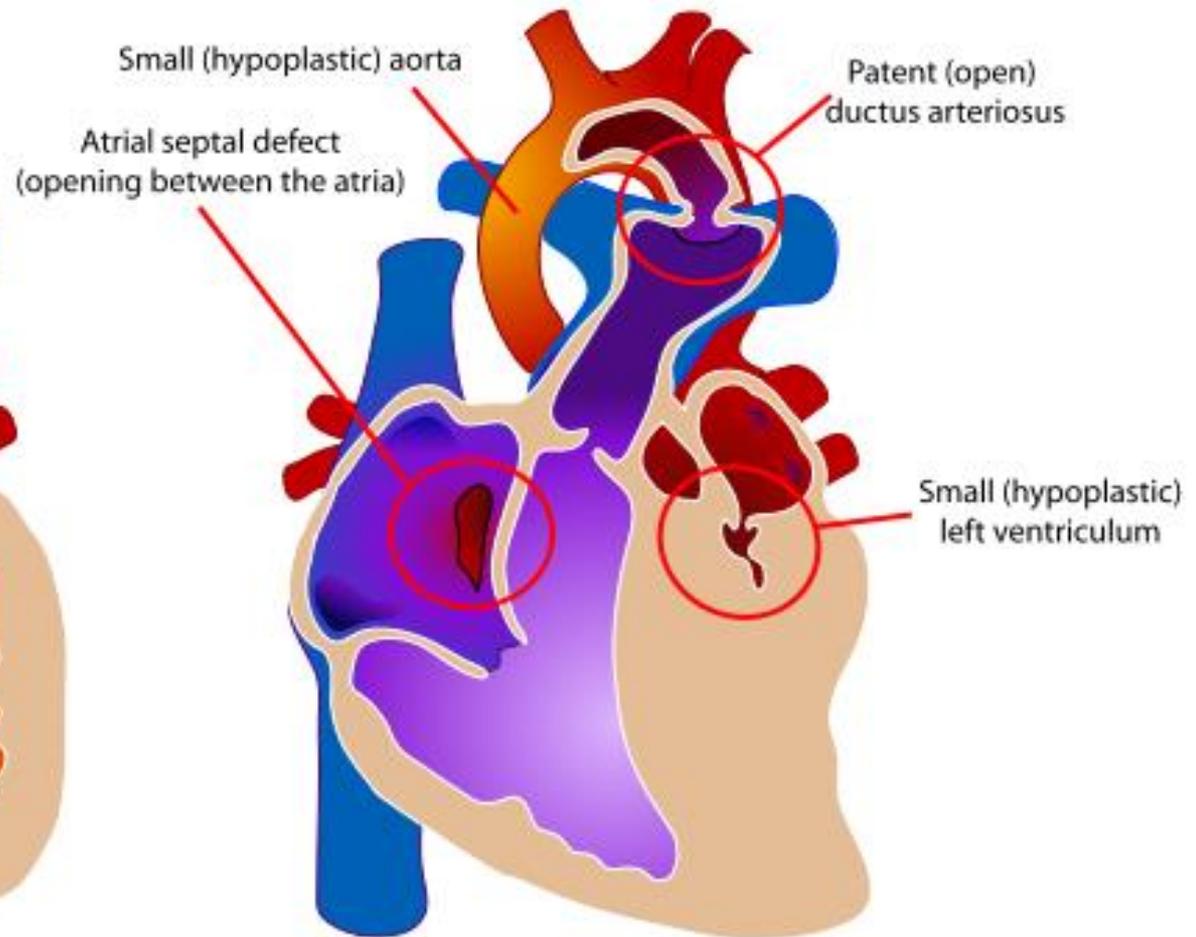
# Hypoplastic Left Heart Syndrome

- 2 in 1,000 live births
- 3% of congenital heart disease
- hypoplasia of the mitral/aortic valves to valvar atresia
- virtually always coarctation of the aorta, and the left ventricle is markedly hypoplastic and dysfunctional
- infants may initially appear well, the only clinical finding may be cyanosis, which may be overlooked
- soft murmur or absence of a murmur because the blood flow typically is not turbulent

Normal heart



Hypoplastic left heart syndrome



- because the ductus arteriosus must supply circulation to the area distal to the area of aortic arch hypoplasia, the oxygen saturation is reduced in the lower body due to the obligate ductal right-to-left shunt of desaturated right heart blood.

# Why not the other options?

- Coarctation of the aorta : absence of a blood pressure discrepancy in the neonate described\*\*\*
- Hypertrophic cardiomyopathy: high-pitched murmur, no oxygen saturation disparity between arm and leg.
- Transposition of the great vessels: it involves the left heart supplying well-saturated blood flow to the pulmonary artery, ductus arteriosus, and lower body, while the desaturated blood from the right heart travels to the aorta and upper body, thus resulting in lower saturation in the upper extremities compared to the lower extremities

## Case 2

A 9 day old male presents to the ER with tachypnea. Antenatal and delivery periods were uncomplicated. He went home with his mother with no concerns on day 2 of life. Tachypnea worsened rapidly over 2 days. In the ER: T37.6C P180 RR85 RUL BP 85/48 LLL BP 65/37. He has a 2/6 systolic murmur in the ULSE. Lower limb pulses are not palpable. Which of the following is NOT appropriate in managing this patient.

A. Take off an ABG

B. Insert a urinary catheter

C. Bolus with N/S 10mL/kg

D. Administer Alprostadil 0.05mcg/kg/min



# Option C again ?

- ABGs monitor for acidosis
- Insertion of urinary catheter allows for monitoring of urine output and renal perfusion
- Alprostadil - prostaglandin E<sub>1</sub> : maintains patency of the ductus arteriosus. The dose is 0.05-0.15 mcg/kg/min.
- The neonate in the vignette is not in shock and thus a bolus is not necessary. Management of CHF involves diuretics and inotropic drugs.

Thank you !



# References

1. <http://www.uptodate.com/contents/congenital-heart-disease-chd-in-the-newborn-presentation-and-screening-for-critical-chd>
2. Question 92 of 2012 PREP Questions and the accompanying explanation
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# Frostbite

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# Question 1

An 8 year old male is brought in by EMS from an icy mountain. He was with his family skiing but was separated from them and alone in the cold for approximately 4 hours. On arrival to the ED, vitals were T 36C, HR 70, RR 20, BP 90/50. On PE, patient is alert and oriented but shivering. You notice large blisters on both of his feet that contain clear fluid surrounded by edema and erythema. The patient has no past medical history. This type of finding would be considered:

# Question 1

- A. First degree frostbite
- B. Second degree frostbite
- C. Third degree frostbite
- D. Fourth degree frostbite

# Question 1

- A. First degree frostbite
- B. **Second degree frostbite**
- C. Third degree frostbite
- D. Fourth degree frostbite

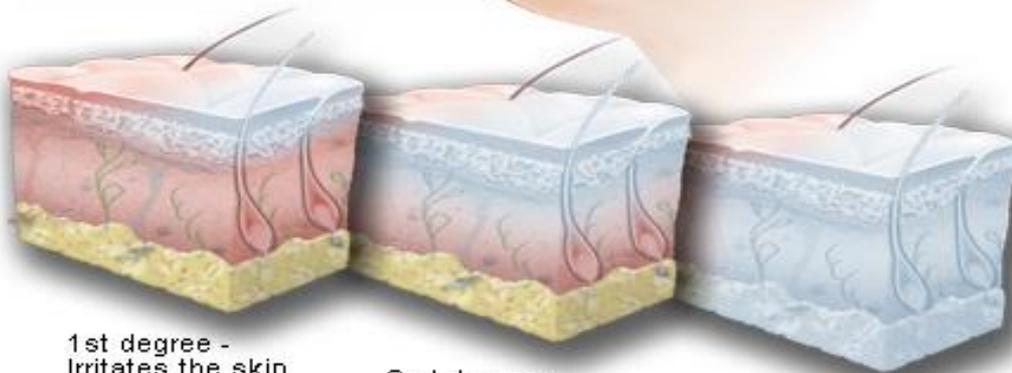
# Question 1-Answers

The answer is B. First-degree frostbite tend to be very superficial and is characterized by a central area of pallor and anesthesia of the skin surrounded by edema. Second-degree frostbite is recognized by large blisters containing clear fluid surrounded by edema and erythema, developing within 24 hours and extending to or nearly to the tips of digits. The blisters can develop an eschar but it eventually sloughs off to reveal healthy granulation tissue with no tissue loss.

# Question 1- Answers

Third-degree frostbite is different from second-degree in that the injury is deeper and the blisters are smaller, hemorrhagic and more proximal. The skin forms a black eschar in one to several weeks. Fourth-degree frostbite, which extends to muscle and bone, involves complete tissue necrosis. Mummification can be seen in 4 to 10 days.

# Frostbite



1st degree -  
Irritates the skin

2nd degree -  
Blisters but has  
no major damage

3rd degree -  
Involves all layers  
of the skin and  
causes permanent  
tissue damage

# Question 2

A 13 year old female is brought in by EMS after being found unconscious by an ice fishing lake in Michigan. It is unclear how long she has been exposed to the cold, but it is estimated that she has been by the fishing lake for about 4 hours. After being given pre-hospital care, the patient's vitals were rectal temperature of 35 degrees celsius, HR 60, RR 15, BP 100/60, O2 sat of 97% on RA. Once airway, breathing and circulation are assessed, preliminary studies are ordered. PE exam reveals a lethargic female with cold, significant for pale extremities with small blisters that appear hemorrhagic on feet and hands. Of the following, which would be the ideal treatments used for frostbite?

# Question 2 - Answers

- A. Rewarming with water heated no higher than 36°C as to prevent a painful warming process
- B. intravenous heparin given along with tetanus prophylaxis
- C. intra-arterial tPA given along with tetanus prophylaxis
- D. debridement of blisters and consult surgery

# Question 2 - Answers

- A. Rewarming with water heated no higher than 36°C as to prevent a painful warming process
- B. intravenous heparin given along with tetanus prophylaxis
- C. intra-arterial tPA given along with tetanus prophylaxis
- D. debridement of blisters and consult surgery

# Question 2--Answers

The answer is C. Several studies have been done in demonstrating the benefit from tPA, which helps prevent vascular thrombosis of the frostbite affected tissue. The recommended dose is a bolus of two to four mg followed by an infusion started 0.5 to 1 mg/hour given via the femoral or brachial artery \* (child dose 0.01-0.06 mg/kg per hour). Repeat angiograms were done every 8 to 12 hours and treatment was continued until perfusion was restored or a time limit of 48 hours was reached. There is no evidence that intravenous heparin alone improves outcomes in frostbite. Tetanus is a reported complication of frostbite, which is why prophylaxis is recommended.

# Questions 2 – Answers

Rewarming is most effectively accomplished by immersing the affected area in water heated to 37° to 39°C, ideally in a whirlpool. Higher temperatures do not warm the injured area any faster and cause the warming process to be much more painful. Dry heat is difficult to regulate and is therefore not used. There is some discord on the management of frostbite-induced blisters. Some believe blister fluid should be drained to eliminate high concentrations of inflammatory mediators.

# Questions 2 – Answers

The other, more conservative approach is to drain, debride, and bandage large non-hemorrhagic bullae that interfere with movement, for instance over joints. *Hemorrhagic bullae* of comparable size and location are drained by aspiration, but are *not debrided*. They usually reflect more significant damage to the microvasculature. **Other minor bullae should be left intact.**

# References

- An Open-Label Study to Evaluate the Safety and Efficacy of Tissue Plasminogen Activator in Treatment of Severe Frostbite—*Journal of Trauma Injury and Critical Care Dec 2005 Volume 59 Issue 6*
- Uptodate: Frostbite
- Uptodate: Treatment of hypothermia in children
- Low-dose tissue plasminogen activator thrombolysis in children. [J Pediatr Hematol Oncol.](#) 2003 May;25(5):379-86.