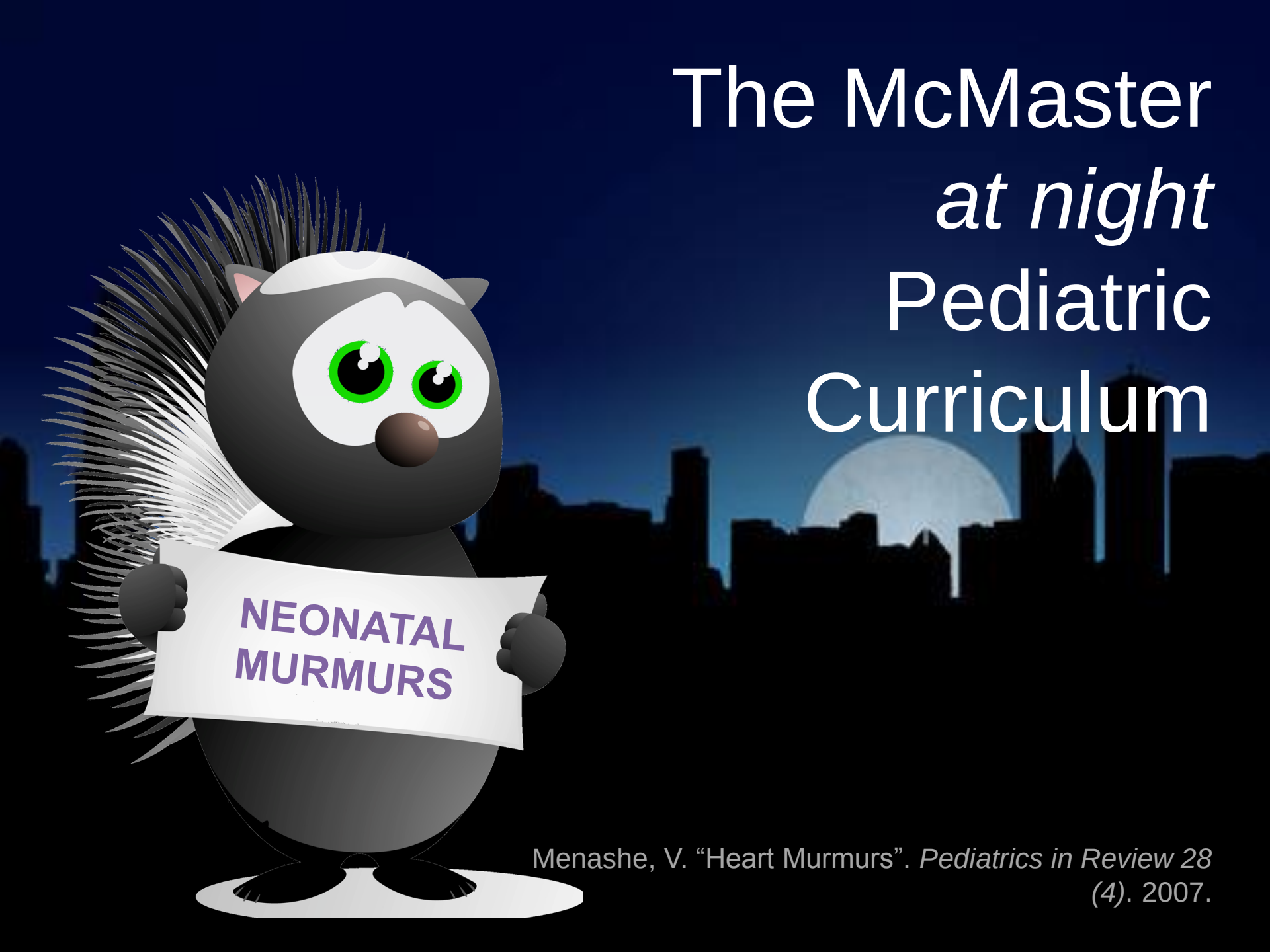
The McMaster *at night* Pediatric Curriculum



Menashe, V. “Heart Murmurs”. *Pediatrics in Review 28 (4)*. 2007.

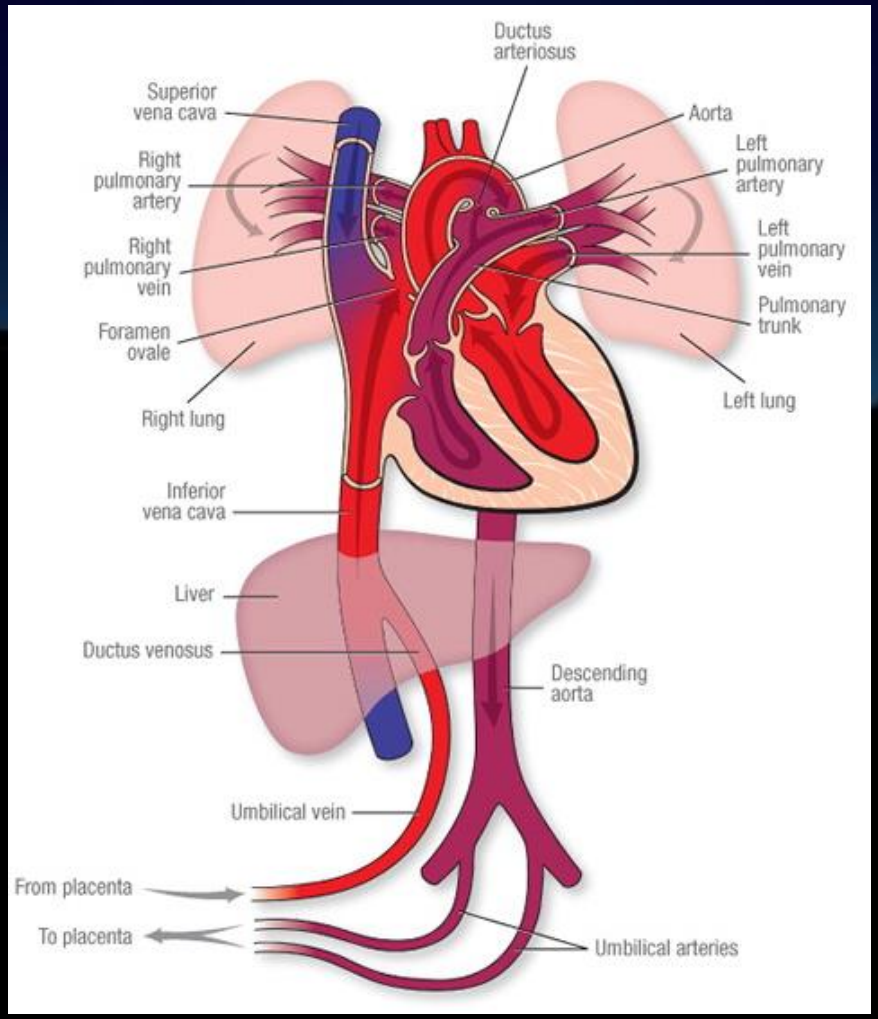
Objectives

- Characterize and describe heart sounds, murmurs, and adventitious sounds
- Distinguish innocent from pathological murmurs that require echocardiography
- Understand the various cyanotic and acyanotic congenital heart defects associated with murmurs
- Complete a thorough history and physical examination in a neonate with a murmur, and recognize indications for emergent treatment

Background

- Congenital heart disease affects 8/1000 live births, with VSD the most common pathology
- CHD may be diagnosed prenatally or during the birth hospitalization, however many cases are asymptomatic in the first days and go undetected
- Most murmurs are non-pathological, but a murmur may be the first or only clue to significant CHD
- Murmurs arise from turbulent blood flow through normal or abnormal heart structures or vessels

The Fetal Circulation



- Features higher right-sided pressure and pulmonary vascular resistance
- The ductus arteriosus normally closes in the first days of life
- The foramen ovale closes after birth as left-sided pressure exceeds right

Background

- Acyanotic heart disease arises when oxygenated blood shunts from left to right across a persistent connection
- Cyanotic heart disease arises when deoxygenated blood shunts from right to left across a persistent connection
- Shock results from outflow tract obstruction
- In duct-dependent lesions, closure can precipitate rapid deterioration to shock, acidosis, and arrest

The Case

- You are called to the well-baby nursery to assess a 2-day old infant girl who is being discharged later this morning
- The family physician detected a murmur and wants to know if the baby needs an echocardiogram
- The baby was born at 38 weeks with a birth weight of 3650g. As you approach, you note that the baby is not in any respiratory distress and is pink aside from hands and feet

History

What would you ask?

History

- Obtain a maternal history including age, serology, prenatal infections, health issues, and medications
- Illnesses of importance include SLE, Sjorgen's, diabetes, CMV, rubella, coxsackie, and herpes
- Teratogens of importance include alcohol, smoking, lithium, antidepressants and anticonvulsants
- A first-degree relative with CHD confers a three-fold risk; also ask about miscarriages, cardiomyopathies, and sudden death

History

- Ask about the results of all prenatal ultrasounds and any prenatal screening
- The birth history includes gestational age, birthweight, delivery complications, APGAR scoring and the need for resuscitation
- When was the murmur first detected, and has it changed?
- How has the infant been feeding, breathing, and sleeping?

Physical Exam

What would you look for?

Physical Exam

- Observe the infant for
  - Respiratory distress: grunting, head bobbing, nasal flaring, tracheal tug, indrawing
  - Colour: central vs peripheral cyanosis, pallor
  - Precordial activity
  - Dysmorphic features

- Obtain a full set of vitals including heart rate, respiratory rate, blood pressure, and temperature

- Blood pressure should be done in all 4 limbs (an average systolic discrepancy of > 10 mmHg in the lower limbs is significant)

Physical Exam

- Compare pre-ductal saturation (right arm) with post-ductal (either leg) to identify right to left shunting across the PDA
- Palpation of the femoral pulses and assessment of peripheral perfusion are essential
- Auscultate lungs for crackles of pulmonary edema
- CHF is unlikely in the immediate neonatal period, but signs include tachycardia, tachypnea, cardiomegaly, and hepatomegaly

Physical Exam

- Murmurs are characterized in terms of:
  - Location and radiation
  - Timing: systolic, diastolic, early, mid, late
  - Contour: holo-, crescendo, decrescendo
  - Pitch: high, low
  - Quality: mechanical, harsh, soft, blowing
  - Intensity:
    - Grade I – barely audible
    - Grade II – audible and constant
    - Grade III – loud without thrill
    - Grade IV – loud with thrill
    - Grade V – stethoscope just touching chest
    - Grade VI – stethoscope off chest

Physical Exam

- Describe the heart sounds
  - S1: AV valve closure
  - S2: aortic and pulmonary valve closure with physiologic splitting during inspiration
  - S3: rapid filling of ventricles (normal in children)
  - S4: stiffness of ventricles (always abnormal)

- Extra sounds include clicks (associated with valvular stenosis or prolapse), and rubs (associated with pericarditis or effusion)

Workup

What would you order?

Workup

- The main consideration is whether or not the exam findings merit echocardiography
- Chest XR detects cardiomegaly and pulmonary congestion, which are non-specific clues to CHD and indications for echo (the cardiac silhouette shape may also have diagnostic value)

Test Your Knowledge

- A full examination reveals a well-looking neonate with heart rate 140, respiratory rate of 40, no distress, acrocyanosis only, and strong femoral pulses. Which of the following characteristics of the murmur would prompt you to arrange for echocardiography?

A. Early systolic onset
B. High-pitched
C. Fixed S2 split
D. Radiation to the back

The Answer

- Innocent murmurs are characterized by the absence of diastolic or pansystolic timing, S2 abnormalities, thrills, extra sounds, and abnormalities on physical exam

  - Fixed S2 occurs in ASD due to increased flow through the pulmonary valve
  - Peripheral pulmonic stenosis is a common non-pathological high-pitched, blowing, systolic ejection murmur that radiates to the back

Differential Diagnosis

Neonatal Murmurs	
Innocent Murmurs	Acyanotic Heart Disease
Peripheral pulmonary stenosis	Atrial septal defect
Still's murmur	Ventricular septal defect
Early patent ductus arteriosus	Patent ductus arteriosus
Cyanotic Heart Disease	Coarctation of the aorta
Truncus arteriosus	Aortic stenosis
Transposition of the great arteries	Pulmonary stenosis
Tricuspid atresia	Bicuspid aortic valve
Tetralogy of Fallot	Mitral regurgitation
Total anomalous pulmonary venous return	Tricuspid regurgitation
Hypoplastic left heart	
Pulmonary atresia or stenosis with intact ventricular septum	

Innocent vs Pathological

Feature	Innocent	Pathological
Intensity	Grade I-II only	Grade I-VI
Timing	Systolic only	Systolic or diastolic
	Early or mid systolic only	Early, mid, or late
Contour	Crescendo-decrescendo only	Crescendo-decrescendo, crescendo, decrescendo, holo-
Location	Lower sternal border, base, supraclavicular	Anywhere
S2	Physiologic splitting	Fixed, wide, single, reversed
Quality	Soft, blowing, vibratory only	Harsh
Extra sounds	None	Possible
Exam	Normal	Shock, cyanosis, pulmonary edema, poor perfusion, absent femorals, dysmorphisms

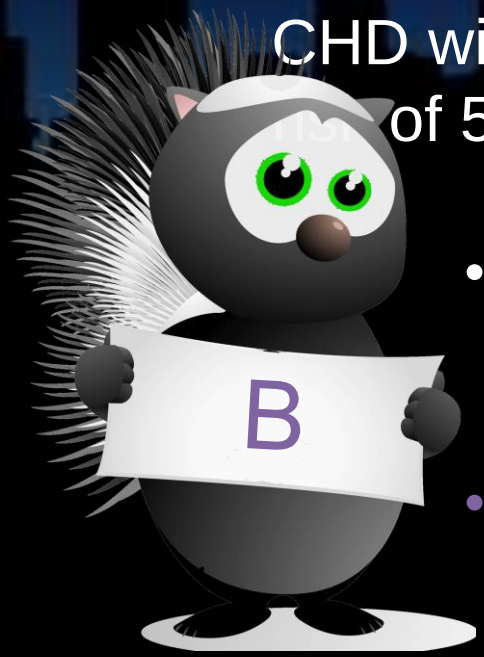
Test Your Knowledge

- You are assessing the 12h-old male infant of a diabetic mother who has been admitted to the level 2 nursery for hypoglycemia. On auscultation you detect a grade III harsh-sounding holosystolic murmur at the lower left sternal border. What do you expect the echo to reveal as the cause of the murmur?

A. Left ventricular hypertrophy
B. Ventricular septal defect
C. Transposition of the great arteries
D. Coarctation of the aorta

The Answer

- Maternal diabetes is associated with cardiomegaly, which can be clinically significant if there is associated left ventricular outflow obstruction
- Maternal diabetes is also a significant risk factor for CHD with a prevalence of 4% and relative risk of 5.0



  - VSD is the most common pathology, but the risk of TGA is increased over 10-fold
  - VSD presents with a harsh holosystolic murmur at the lower left sternal border

Innocent Murmurs

- PPS
  - Mid-systolic, blowing, high-pitched, low-intensity murmur best heard at the base with radiation to the back
  - Represents flow through the acute angle of branch pulmonary arteries
- Still's murmur
  - Low-pitched, low-intensity, vibratory systolic ejection murmur best heard at the lower left sternal border
  - Represents pulmonic valve leaflet vibration
  - More common in children than newborns

Acyanotic Heart Disease

Lesion	Murmur	XR
ASD	Fixed S2 split Midsystolic ejection murmur Best heard at the base	Enlarged RA, RV ↑ pulmonary flow if large
VSD	Variable S2 splitting Harsh holosystolic murmur Best heard at left lower sternal border Pitch depends on size Intensity varies inversely with size	Enlarged LA, LV ↑ pulmonary flow if mod-large
PDA	Continuous machinery murmur Grade I-IV depending on size Left infraclavicular region	Normal if small Prominent PA and aortic notch if moderate ↑ pulmonary flow and cardiomegaly if large
Coarctation	Systolic ejection murmur Best heard at the apex Systolic click if BAV also present	Cardiomegaly and ↑ pulmonary flow if CHF Rib notching in children

ASD

- Accounts for 13% of CHD with 1.6/1000 prevalence
- The degree of left-to-right shunting depends on the size of the defect, pressure gradient, and relative compliance of the left and right ventricles
- Blood flow through the right heart, pulmonary vasculature and left atrium is increased
- Most lesions remain asymptomatic for years
- The majority of lesions < 7mm close spontaneously

ASD

- For the remainder, over time, pulmonary hypertension and right-to-left shunting develop (Eisenmenger syndrome)

VSD

- Accounts for 50% of CHD with 4.2/1000 prevalence
- The degree of left-to-right shunting depends on the size of the defect
- Blood flow through the pulmonary vasculature, left atrium, and left ventricle is increased
- Over time, pulmonary hypertension and right-to-left shunting develops (Eisenmenger syndrome)
- 75% of small defects undergo spontaneous closure and those that persist are asymptomatic and benign

VSD

- The prognosis for moderate defects ranges from spontaneous closure to symptoms of high-output failure in the first weeks of life (surgical intervention is indicated when RV pressure exceeds 50% of LV pressure)

- Large VSDs equalize pressure between the ventricles and rarely close spontaneously (surgery is required in the first year of life)

PDA

- Risk factors for PDA include prematurity, maternal rubella infection, RDS, and Down syndrome
- Depending on size, PDA may be asymptomatic but classically presents with bounding pulses, active precordium, tachypnea and poor growth
- A continuous machine-like murmur or systolic ejection murmur is usually present
- Symptomatic PDAs are treated with IV indomethacin or ibuprofen; refractory cases can be surgically ligated

Cyanotic Heart Disease

Lesion	Description	Murmur	XR
Truncus arteriosus	Single great vessel arises from heart	Single loud S2 Ejection click VSD murmur present	Cardiomegaly ↑ pulmonary flow
Transposition of great arteries	Aorta arises from RV, while PA arises from LV	Not a prominent feature, unless VSD also present	Egg on a string ↑ pulmonary flow
Tricuspid atresia	Atresia, stenosis or reversal of tricuspid valve	Widely split S1, S2 Prominent S3, S4 Systolic TR murmur	Cardiomegaly ↓ pulmonary flow
Tetralogy of Fallot	Overriding aorta, RVH, PS, VSD	Single S2, harsh systolic ejection murmur of PS	Boot ↓ pulmonary flow
TAVPR	All 4 pulmonary veins drain into left circulation	Fixed split S2 Systolic murmur Diastolic rumble	Snowman ↑ pulmonary flow
Hypoplastic left heart	Underdeveloped LV, MV, AV	Single S2 No murmur	Variable and non-specific

Test Your Knowledge

- You are called urgently to assess a 3-day-old who has been feeding poorly all day and has increasing work of breathing. You discover a centrally-cyanosed baby who is grunting and indrawing with a HR of 190, RR of 80, preductal saturation of 72% and postductal saturation of 80% despite 50% FiO_2. There is no murmur. What intervention is most likely to influence the outcome?

A. Increase the FiO2 to 100%
B. Rapid sequence intubation
C. Stat chest XR
D. IV alprostadil

The Answer

- Any 2-7 day old infant presenting with cyanosis or shock should raise strong suspicion of a duct-dependent cardiac lesion

- The lesion described is TGA, which often features no murmur and is the only instance where postductal saturations are higher

  - Prompt initiation of IV prostaglandin maintains the PDA, temporizing the infant until emergent intervention

  - Increasing the FiO_2 will have no effect

Duct-Dependent Lesions

- Lesions with duct-dependent pulmonary circulation (pulmonary flow is supplied by aorta) usually present with cyanosis
  - Pulmonary atresia/stenosis
  - Severe tetralogy of Fallot
  - Tricuspid anomalies

- Lesions with duct-dependent systemic circulation (systemic circulation is supplied by pulmonary artery) usually present with shock
  - Hypoplastic left heart syndrome
  - Coarctation or severe aortic stenosis

Summary

- Neonatal murmurs are very common, and while most are non-pathological, murmur may be the first presentation of congenital heart disease
- Characterization of the murmur along with complete history and exam will direct the most at-risk patients to echocardiography
- Many critical cardiac lesions are asymptomatic in the first days of life, highlighting the importance of thorough exam, close follow-up, and early recognition of critical events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