Innocent Murmurs

- Diastolic murmurs are never innocent
- Innocent murmurs are present in at least 50% of normal children
  - Still’s murmurs: low pitched, vibratory, systolic ejection, increases with the supine position.
  - Venous hum: continuous murmur in supraclavicular region, reduces on lying down or with pressure on neck.
Upon physical examination of a 3 year old girl who is new to the practice, you note a continuous grade 2 to grade 3 murmur at the upper right sternal border while she is sitting. In the supine position, you note only a grade 2 low pitched systolic murmur at the apex. Measurements of BP, pulses and precordial palpations as well as the auscultation is normal. Of the following, the MOST appropriate next step is to:

1. Reassure the parents about the benign prognosis
2. Request a cardiology consultation
3. Request chest radiography
4. Request echocardiography
5. Request electrocardiography

Congenital Heart Disease - Structural

- **PINK**
  - Shunts (L to R):
  - ASD
  - VSD
  - PDA

- **BLUE**
  - TOF
  - TGA
  - Tricuspid atresia
  - Truncus
  - TAPVR
  - Ebstein’s
  - Single ventricle

Normal Cardiac Pressures

<table>
<thead>
<tr>
<th></th>
<th>RA</th>
<th>LA</th>
<th>RV</th>
<th>LV</th>
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<tr>
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<td>25/15</td>
<td>120/80</td>
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</table>
ATRIAL SEPTAL DEFECT

ATRIAL SEPTAL DEFECTS (ASD)

- Three types exist: primum, secundum and sinus venosus
- The most common is the secundum type
- Symptoms: None in childhood, arrhythmias in the 3rd decade

ASD- cont...

- Clinical signs include a 2-3/6 SEM at the ULSB and a fixed wide split S2
- A large ASD causes right ventricular enlargement
- EKG: RAD and IRBBB
ASD - cont...

- ECHO: Diagnostic
- Natural History: Arrhythmias and pulmonary obstructive vascular disease in the 3rd and 4th decade.
- Treatment: Surgical vs. transcatheter closure
VENTRICULAR SEPTAL DEFECT

VENTRICULAR SEPTAL DEFECTS (VSD)
- This is the most common form of CHD
- The VSDs are subdivided according to the part of the septum they occur in: Muscular, perimembranous, inlet, outlet
- A large VSD causes left ventricular enlargement
- With a small VSD there is normal growth and development

VSD - cont.....
- With a large defect there may be CHF (usually at 6-8 weeks), pulmonary infections and delayed growth
- Clinical signs: Loud 4-5/6, harsh holosystolic murmur, middiastolic rumble and a loud P2
EKG: LVH or BVH

ECHO: Diagnostic

Natural history: Small VSDs close spontaneously depending on the site.

Unrepaired the large defects may lead to Eisenmenger’s syndrome.

Large VSDs are closed surgically by 6 months of age.

Diuretics, digoxin and afterload reducing agents are used prior to surgery - if needed.
ENOCARDIAL CUSHION DEFECTS

AVSD - cont...

- 1/3rd of babies with this have Down syndrome
- EKG: Characteristic with a superior left axis.
- Echo: Confirmatory
- Management: Anticongestive medications and surgery at 4-8 months of age.
PATENT DUCTUS ARTERIOSUS (PDA)

- It is a connection between the aorta and the pulmonary artery.
- Very common in preterm babies.
- Usually closes in the first 2 weeks of life.

PDA - cont.....

- Symptoms: a) None if small
  - b) If large can cause CHF at 6-8 weeks in a term infant
  - c) In a preterm baby increasing respiratory support usually occurs after day 3 of life.
PDA

- Signs: Systolic murmur in a newborn and a continuous “train in a tunnel” murmur in an older child. Best heard below the left clavicle.
- A large PDA causes LA and LV enlargement.
- Treatment: Preterm vs. term baby.

PDA - cont...

- In a preterm it can be closed medically using indomethacin.
- In a term baby if still open at 3 months of age then coil closure by cardiac catheterization is the method of choice.

A 3 month old girl who has Down syndrome exhibits poor weight gain, tachypnea and a low pitched grade 2 murmur. Chest radiography reveals cardiomegaly and increased pulmonary vascularity. EKG documents RVH and a superior frontal plane QRS. Of the following, the MOST likely diagnosis is:

A. coarctation of the aorta
B. complete atrioventricular septal defect
C. patent ductus arteriosus
D. Perimembranous VSD
E. secundum ASD
A 5 day old infant born at 31 weeks gestation is on ventilatory support. He has a grade 2 holosystolic murmur that extends past the second heart sound. Pulses are bounding. Precordial palpation is hyperdynamic. Concentrations of hemoglobin, electrolytes and creatinine are normal. Of the following the most appropriate INITIAL management is to:

A. administer furosemide intravenously
B. administer indomethacin intravenously
C. perform an echocardiogram
D. defer intervention because spontaneous closure is likely
E. obtain a surgical consultation for ligation of the ductus.

You are evaluating a newborn 6 hours after his birth. Labor and delivery were uncomplicated, but amniocentesis performed during the pregnancy revealed trisomy 21. Fetal echocardiography at 20 weeks’ gestation showed normal findings. The infant currently is sleeping and is well-perfused, with a heart rate of 140 beats/min and no audible murmurs. His physical features are consistent with Down syndrome. Of the following, the MOST appropriate diagnostic study to perform is:

1. barium swallow
2. cervical spine radiography
3. Echocardiography
4. head ultrasonography
5. radiography of the abdomen

A term newborn has tachypnea, rales, tachycardia, audible gallop and diminished arm and leg pulses. Echocardiography shows enlargement of both ventricular chambers with good systolic function and no congenital heart disease. Of the following, the MOST likely diagnosis is:

A. Carnitine deficiency
B. Hyperthyroidism
C. Hypoglycemia
D. intracranial arteriovenous malformation
E. pheochromocytoma
A term newborn has tachypnea, rales, tachycardia, audible gallop and diminished arm and leg pulses. Echocardiography shows enlargement of both ventricular chambers with good systolic function and no congenital heart disease. Of the following, the MOST likely diagnosis is:

A. Carnitine deficiency  
B. hyperthyroidism  
C. hypoglycemia  
D. intracranial arteriovenous malformation  
E. pheochromocytoma

COARCTATION OF THE AORTA

- More common in males  
- Almost always juxtaductal  
- 85% of children with CoA have a bicuspid aortic valve.
CoA - cont....

- Symptoms and Signs:
  - SEVERE: Shock
  - MODERATE: CHF,
  - MILD: Headaches, leg claudication
- Decreased femoral pulses are an important sign esp. in neonates.
- BP lower in the lower limbs

CoA - cont....

- ECHO: Diagnostic
- Treatment: For an infant in shock - PGE1 immediately.
- Surgical vs. transcatheter repair.

Hypoplastic Left Heart Syndrome

- Varying degrees of left heart hypoplasia at multiple levels
- Babies present in cardiogenic SHOCK once the ductus closes.
- Immediate treatment is PGE1 intravenously as an infusion.
Hypoplastic Left Heart syndrome

- Surgical Treatment:
  - Norwood at birth
  - Glenn at 4-8 mnths
  - Fontan at 2-4 years

A 7-month-old female has undergone the second stage of surgical palliation (Glenn operation) for hypoplastic left heart syndrome. She was discharged from the hospital 1 week ago, and her mother brings her to the office because of irritability that began this morning. On physical examination, the infant is awake and irritable, with a heart rate of 150 beats/min and a respiratory rate of 50 breaths/min. She has cyanosis of the face and mucosal surfaces and swelling of the arms and head.

Of the following, the BEST explanation for this patient’s clinical presentation is

- Polycythemia
- Postpericardiotomy syndrome
- Protein-losing enteropathy
- Superior vena cava syndrome
- Thoracic duct injury

Pulmonic/ Aortic Stenosis
Stenosis

Pulmonic
- This may be at the valve, subvalvar or supravalvar.
- Symptoms: None in mild or moderate stenosis. Cyanosis is seen only with critical PS.
- Signs: Ejection click and a harsh SEM, at the ULSB.
- ECHO: Diagnostic
- Treatment: Ballooning

Aortic
- Stenosis possible at the valve, subvalvar or supravalvar.
- This is a more significant and a dangerous lesion compared to PS.
- More common in males.
- Valvar AS is usually associated with a bicuspid aortic valve.
- Treatment: Ballooning

AS
- A type of subvalvar AS is also called HCM which is the commonest cause of sudden death in children
- Symptoms:
  - Mild: None
  - Moderate to severe: Chest pain, fatigability, syncope.

HYPERTROPHIC CARDIOMYOPATHY
A 3 day old girl is found unconscious in her crib and is brought to the ED. Findings include: tachypnea, tachycardia, pallor; poor capillary refill; hepatomegaly; cardiomegaly with increased pulmonary vascular markings; hemoglobin concentration 17 gm/dl; and hematocrit, 51%. Of the following, the cardiogenic shock in this girl MOST likely is due to:

A. critical aortic stenosis
B. erythroblastosis fetalis
C. severe hypovolemia
D. ventricular septal defect

A 6 hour-old infant has increasing pallor, tachypnea and respiratory distress. Physical examination reveals an enlarged liver, a gallop rhythm, poor pulses in the upper extremities and absent pulses in the lower extremities. In addition to treating the infant for sepsis, the most appropriate INITIAL management is to administer:

1. a dopamine infusion
2. a loading dose of digoxin
3. a 25% glucose and water solution
4. Furosemide
5. prostaglandin E1.
There has to be a RIGHT to LEFT shunt to cause cyanosis

Tetralogy of Fallot

- Most common cyanotic heart disease.
- The four abnormalities include:
  - Pulmonary stenosis
  - RVH
  - VSD
  - Overriding Aorta
- Signs include cyanosis, murmur, squatting and spells.
A “tet” spell consists of rapid breathing and increased cyanosis. Any event like crying or increased physical activity can initiate the spell.

Treatment includes:
- holding the baby in a knee chest position
- Morphine
- Oxygen, beta blocker, general anesthesia,
Transposition of the great Arteries

- The aorta arises from the right ventricle and the pulmonary artery from the left.
- The mixing of the blood occurs at the PFO and the PDA.
- The signs include cyanosis and cardiomegaly.
  - **Reverse differential cyanosis**!
- There may be no murmur.
- An echocardiogram is diagnostic.
The mother of a 5 month old girl reports that following a feeding, the child began to breathe deeply, became increasingly blue and then lost consciousness. After being held briefly, the infant regained her usual color and became alert. Physical examination reveals a harsh murmur. Of the following the MOST likely diagnosis is:

A. aortic stenosis
B. coarctation of the aorta
C. myocarditis
D. tetralogy of Fallot
E. ventricular septal defect

You are called at 3 AM from the nursery where 36 hour old BB Bleu is noticed to be cyanotic. The nurses report that he had been feeding well and appeared healthy with Apgar scores of 9/9. Until tonight he appeared pink. They report no significant tachypnea. You order a chest X-Ray and pulse oximetry to be done while you rush to the hospital. On arrival the pulse oximetry indicated O2 saturation of 55% and the X-ray shows no increase in pulmonary vascular markings or infiltrate. The next MOST appropriate intervention is to:

A. obtain a stat EKG to evaluate for SVT
B. intubate the infant and place on 100% O2.
C. start IV prostaglandin infusion at 0.05-0.2 mcg/kg/min
D. start nitric oxide at 40ppm inspired to reduce pulmonary vascular resistance
E. arrange for transfer to a facility capable of ECMO.

Following an uncomplicated delivery, a 3.7 kg term infant develops cyanosis in the first hour of life. Findings at 3 hours of age include: cyanosis; heart rate, 140 beats/min; respiratory rate, 56/min; no heart murmurs; pulse oximetry in room air, 70% saturation in the right hand and 75% in the foot; in 100% FIO2 via head-hood oxygen, saturation increases to 90% in the foot; chest radiography, normal. These findings are most consistent with:

1. Primary pulmonary Hypertension of the newborn
2. pulmonary valve atresia
3. transient tachypnea of the newborn
4. transposition of the great arteries
5. truncus arteriosus
At 60 minutes of age, a term 3.3-kg female infant appears cyanotic but is otherwise well. Her oxygen saturation is 79%, she has widespread cyanosis, and you can hear a faint low-pitched murmur diffusely across the chest. The remainder of findings on her physical examination are within normal limits. After placing her on nasal cannula oxygen at 2 L/min, you note no change in saturation. Of the following, the MOST likely cause of this child’s findings is:

A. Anemia  
B. Hypoplastic left heart syndrome  
C. Neonatal sepsis  
D. Retained fetal lung liquid syndrome  
E. Tracheoesophageal fistula

An infant with severe cyanosis presents. For which of the following conditions would balloon atrial septostomy be helpful?

A. Tetralogy of Fallot  
B. Transposition of the Great arteries  
C. Truncus Arteriosus  
D. Anomalous pulmonary venous return  
E. Large VSD

Congestive Cardiac Failure

- Tachycardia  
- Tachypnea  
- Hepatomegaly  
- Cardiomegaly, murmur, HR too fast/slow  
- FAILURE TO THRIVE

CHD  
- 2 months: VSD, PDA  
- Within 1st month: Coarctation, AS, HLHS  
- Neonatal period: Truncus Arteriosus  

Normal heart  
- Myocarditis
In addition to irritability, sweating and difficulty breathing with feeding, the symptom that is MOST indicative of congestive cardiac failure in a 3 week old infant is:

A. ascitis  
B. cough  
C. cyanosis  
D. diminished feeding volume  
E. pretibial edema

A term infant is born with a large ventricular septal defect. At what age is this infant MOST likely to first demonstrate clinical findings of congestive cardiac failure?

1. 2 days  
2. 2 weeks  
3. 2 months  
4. 6 months  
5. 12 months

Rhythm Abnormalities

- Ectopic beats: premature atrial, ventricular  
  - Benign if they disappear with exercise  
  - Seen in the neonatal and adolescent age groups
- Atrial Flutter, fibrillation
- SVT
- VT  
  - Electrolyte Imbalances  
  - TOF  
  - HCM, Long QT syndrome
- AV block
An 8 year old previously healthy boy presents for a school physical. He is active and has no symptoms. On exam. He appears well. His pulse noted by the nurse to be 80 but with periods of bradycardia to 60 and then followed by more rapid rates of 90/min. No other abnormalities are noted.

His EKG:

Most common cause of irregular rhythm in children – SINUS ARRHYTHMIA – BENIGN!!!

Irregular rhythm in a newborn baby- Premature atrial contractions – BENIGN!!!
Irregular rhythm incidentally noted in an adolescent - Ventricular Premature beats which decrease with exercise – BENIGN!!!

**SVT**

- Rate above 230/min.
- Tachycardia – most likely SVT
  - Narrow complex tachycardia
  - WPW is the most common cause of reentry tachycardia in children
Treatment of SVT

- Hemodynamically stable:
  - Vagal maneuvers
  - Adenosine
  - Verapamil in children over 1 year
  - Amiodarone

- Hemodynamically unstable:
  - DC cardioversion

- Chronic M/t:
  - Drugs: Beta blockers, digoxin
  - Radiofrequency ablation

A 1 year old child is brought to the ER because his parents thought his heart was pounding as they were putting him to bed. EKG reveals a HR of 300/min that spontaneously converts to a sinus rate of 100/min. The parents estimate that the tachycardia lasted 20 minutes; the child was asymptomatic throughout. Of the following the MOST appropriate management of this child is:

A. administration of a beta blocker
B. administration of digoxin
C. administration of procainamide
D. administration of verapamil
E. observation without drug therapy
A 4 week old infant appears in your ED with a history of irritability, increased respiratory rate and poor feeding. On physical examination the child is diaphoretic with decreased perfusion and tachypneic but still alert. You notice no murmur but the monitor indicates a HR of 280 bpm. All but one of the following are appropriate:

A. obtain a 12 lead EKG
B. give verapamil 0.1 mg/kg push slowly
C. give adenosine 100 mcg/kg rapid push
D. fill a bag with ice and apply to infants face
E. pass an esophageal probe and pace the heart 20 bpm faster than the tachycardia

Atrial Flutter/ Fibrillation

- Seen in two groups
  - Newborns: After o/t BENIGN!!
  - After extensive atrial surgery such as Fontan op, atrial switch for TGA etc.
- Treatment: DC Cardioversion, AV blocking meds

AV BLOCK

- **First Degree** – Prolonged PR interval
  - Rheumatic fever, ASD, PDA
- **Second Degree**
  - Type I: Varying PR intervals and dropped beat, Wenkebach
  - Type II: 2 or more than 2 :1 block
- **Third Degree**:
  - Surgical, Lyme Disease
  - Mom with SLE
4. SYNCOPE

- Brief loss of consciousness with rapid recovery
- Seen in adolescents and in toddlers
- 20-50% of adolescents experience at least one episode of syncope
  - Most cases benign
  - Vasovagal syncope is the most common type in adolescents
- Typical history, normal EKG
BENIGN SYNCOPE

- Vasovagal
- Orthostatic hypotension
- Hyperventilation
- Breath holding spells

DANGER SYMPTOMS

- Syncope especially with EXERTION or EXCITEMENT - anger, fear, startle
- Cardiac arrest with exercise or excitement

Sudden Death in Young Athletes
Commotio Cordis

- Young children
- Baseball, football, ice hockey
- Force of blow is not unusually hard
- R on T phenomenon
- Prevention: ? softer balls, ? protective clothing,
- Role of automated External defibrillator

Long QT Syndrome

- Disorder of the electrical activity of the heart
- Involves repolarization
- Characterized by QT prolongation
- Pts. are susceptible to sudden death due to Torsade de pointes
- Syncope typically occurs with a startle or exertion
- can be inherited or acquired
Special situations where the QT should **ALWAYS** be measured:

- Syncope
- Seizures
- Congenital Deafness
- Near SIDS
A 5 year old girl is very excited following a ride on the ferris wheel. In the midst of her excitement she suddenly loses consciousness and falls to the ground. Paramedics on the scene document ventricular tachycardia. Family history reveals a maternal uncle who died suddenly at 16 years of age. Following treatment of the ventricular tachycardia, an electrocardiogram most likely will demonstrate:

A. corrected QT interval of 0.52 sec  
B. P wave axis of –30 degrees  
C. PR interval of 0.81 sec  
D. QRS axis of –15 degrees  
E. QRS interval of 0.12 seconds

A 12 year old boy underwent repair for tetralogy of Fallot at 9 months of age. Last month, routine follow up echocardiography revealed no residual shunts; moderate right ventricle enlargement; a 50 mm Hg gradient from the right ventricle to the main pulmonary artery; and normal LV size and function. Today he is dizzy and had a near syncopal episode in gym class. The MOST likely cause for his symptoms is:

A. left ventricular failure  
B. physical deconditioning  
C. pulmonary hypertension  
D. right ventricular failure  
E. ventricular arrhythmia

Julie, an otherwise healthy 9 year old comes to the ED because she “passed out”. After asking questions and examining the patient all but one of the following reassures you that she has vasovagal syncope which is a relatively benign cause of syncope in children.

A. Julie was standing in line waiting to see “Harry Potter and the Deathly Hallows” when she passed out.  
B. she fainted once before when she had a blood test  
C. after falling to the ground she came to quickly and remembered feeling warm and dizzy  
D. Julie was lying on a sofa watching TV when a door slammed and she suddenly became unresponsive  
E. S1 and S2 were normal and no murmurs were noted
A 14-year-old girl falls during a race. She is unconscious, cyanotic and has no pulse but spontaneously revives within seconds. Both patient and family histories are benign. Results of the physical examination, chest radiography, EKG, echocardiography, EEG and an exercise ECG during a treadmill stress test are normal. The most appropriate NEXT step in management is:

A. order a 30 day looping event recorder
B. perform cardiac catheterization studies
C. Perform 24 hour ambulatory ECG monitoring
D. perform tilt table testing
E. reassure the family that cardiac etiologies have been excluded

A 13-year-old boy wishes to participate in competitive sports. His father died suddenly at age 28 years, and hypertrophic cardiomyopathy was found on autopsy. Of the following, the MOST helpful test for assessing the boy’s risk is:

A. echocardiography
B. electrocardiography
C. exercise myocardial perfusion scintigraphy
D. Genetic testing for myosin chain mutations
E. Genetic testing for troponin mutations

While running sprints during conditioning exercises for soccer, a 17-year-old girl suddenly collapses. Her coach reports that she “woke up after 30–45 seconds” and was immediately oriented and appeared in no acute distress. Upon arrival to the emergency department, her vital signs are stable. Physical examination is unremarkable. She takes no daily medications and denies chronic medical problems. A urine drug screen is negative. An ECG reveals a right bundle branch block and ST-segment elevation in leads V1–V3.

A. WPW syndrome
B. Primary pulmonary hypertension
C. Brugada Syndrome
D. Aberrant left coronary artery
E. Hypertrophic Cardiomyopathy
During a preparticipation sports physical for basketball, a 16-year-old male is noted to have a midsystolic click on cardiac exam. He wears glasses, is tall for his age, has a reduced upper segment-to-lower segment ratio, and has mild scoliosis.

In addition to mitral valve prolapse, which of the following is most likely to be identified during an echocardiogram in this patient?

A. Bicuspid aortic valve
B. Pulmonic stenosis
C. Atrial septal defect
D. Dilated aortic root
E. Asymmetric septal hypertrophy

You are examining a 6 year old new to your practice. He is on a “blue pill” for behavioural issues. He has a round face, flattened bridge of nose, long upper lip. He is obviously intellectually disabled but is very personable and happy. Which cardiac lesion does this child likely have?

A. Supravalvar AS
B. Pulmonary stenosis
C. ASD
D. VSD
E. Atrioventricular septal defect
Congenital Heart Disease-Etiology-Environmental Factors/Toxins

- Lithium: Ebstein’s anomaly
- Ethanol: ASD, VSD (Fetal Alcohol Syndrome)
- Anticonvulsants: PS, AS, TOF
- Retinoic Acid: Transposition
- Rubella: PDA, PPS
- Coxsackie B: Neonatal myocarditis
- Maternal Diabetes: HCM, TGA
- Maternal Lupus: Complete heart block
- PKU: VSD, ASD, complex CHD

A 3-year-old girl presents for her first visit after being adopted from an orphanage in the Ukraine. Her height and weight are at the 10th percentile. Head circumference is below the 3rd percentile. On examination, several dysmorphic facial features are evident, including short palpebral fissures; mild bilateral ptosis; midface hypoplasia; a long, thin philtrum; and a thin upper lip. She was normotensive, and there was no gradient noted between upper and lower extremity blood pressures. Common cardiac finding?

A. An ejection click heard best at the left upper sternal border immediately following the 1st heart sound during expiration
B. A late 2/6 systolic murmur preceded by a click
C.Bounding peripheral arterial pulses
D. A harsh 3/6 (holosystolic) murmur heard best over the lower left sternal border
E. An apical diastolic murmur

PERICARDITIS

- Follows a viral URI
- Sharp chest pain, retrosternal, difficulty in deep inspiration
- Pt. Resists lying down
- Pain worsened by pressure over the sternum
- Friction rub, pulsus paradoxus
- EKG is diagnostic
Pericarditis

MECHANISM OF PULSUS PARADOXUS

- Increased RV flow
- Decreased RV flow and pressure
- Increased return flow and pressure
- Decreased return flow and pressure

PERICARDITIS - EKG

TREATMENT

- Reassurance
- NSAIDS
- Occasional pericardial tap, window
- Postpericardiotomy Syndrome: 2 weeks after surgery
Infective Endocarditis

- The endocardium is a deterrent to adhesion by platelets and organisms.
- The denuded endothelium is a site for platelet adhesion and subsequent vegetation growth.
- The “Low pressure sink” is the site for vegetations.
- Polycythemia

IE- Lab. Tests

- BLOOD CULTURES
- Echo

Prevention of Infective Endocarditis
Guidelines From the American Heart Association

A Guideline From the American Heart Association
Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group

Circulation 2007;116;1736-1754;
Conclusions

(1) Only an extremely small number of cases of infective endocarditis might be prevented by antibiotic prophylaxis for dental procedures even if such prophylactic therapy were 100% effective.

(2) IE prophylaxis for dental procedures is reasonable only for patients with underlying cardiac conditions associated with the highest risk of adverse outcome from infective endocarditis.

(3) Administration of antibiotics solely to prevent endocarditis is not recommended for patients who undergo a genitourinary or gastrointestinal tract procedure.

- Prosthetic cardiac valves or prosthetic material used for cardiac valve repair
- Previous IE
- Congenital heart disease (CHD)*
  - Unrepaired cyanotic CHD, including palliative shunts and conduits
  - Completely repaired congenital heart defect with prosthetic material or device, whether placed by surgery or by catheter intervention, during the first 6 months after the procedure
  - Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
- Cardiac transplantation recipients who develop cardiac valvulopathy
Which of the following are the most common organisms to cause infective endocarditis in children?

A. *Streptococcus pneumoniae* and non-typable *Haemophilus influenzae*
B. *Viridans* group streptococci and *Staphylococcus aureus*
C. *Streptococcus pyogenes* and *Escherichia coli*
D. Coagulase-negative *Staphylococcus* and *Streptococcus pyogenes*
E. Group B *Streptococcus* and coagulase-negative *Staphylococcus*
**Myocarditis - signs and symptoms**

- **DILATED** heart
- **Inflamed** Myocardium and conduction system

<table>
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<tr>
<th>Sinus</th>
<th>TACHYCARDIA</th>
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<tbody>
<tr>
<td>CHF</td>
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</tbody>
</table>

**Arrhythmias**

**Causes**
- Viral
- Bacterial

**Symptoms**
- Chest pain
- C. pain, irr. beats
- Fever

**Signs**
- Rub
- Tachycardia
- Fever

**Tests**
- EKG, echo
- CXR, echo
- B. Culture

**Treatment**
- NSAIDS
- IVIG
- Antibiotics

**Course**
- Benign
- Can be fatal
- Insidious

**Jones’ Modified Criteria**

<table>
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<tr>
<th>Requirement for Diagnosis</th>
<th>Major Criteria</th>
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<tr>
<td>a) Two major criteria or</td>
<td>Carditis</td>
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<tr>
<td>b) One major plus two</td>
<td>Arthritis</td>
</tr>
<tr>
<td>minor criteria</td>
<td>Chorea</td>
</tr>
<tr>
<td>c) Plus evidence of</td>
<td>Erythema marginatum</td>
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<tr>
<td>previous streptococcal</td>
<td>Subcutaneous nodules</td>
</tr>
<tr>
<td>infection:</td>
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<tr>
<td>- Recent scarlet fever</td>
<td>Previous rheumatic fever</td>
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<td>or</td>
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<td>- Raised antistreptolysin O</td>
<td>Fever</td>
</tr>
<tr>
<td>(ASO) titer or</td>
<td>Acute phase reactants</td>
</tr>
<tr>
<td>- Positive throat culture</td>
<td>First-degree atrioventricular block</td>
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</table>
Rheumatic Carditis
- Present in 50% cases
- “Sleeping” tachycardia is an early sign
- Mitral and aortic valves most commonly involved

Rheumatic Arthritis
- Most common manifestation
- Pain, swelling and erythema
- Resolves within 1 week

RF - Treatment and Prevention
- Benzathine penicillin 1.2 mega units IM
- Aspirin 75-100 mg/kg for 6-8 weeks
- Steroids for severe carditis
- Digoxin, diuretics
- Prevention with BP q 4 weeks.

Two weeks after a nonspecific upper respiratory infection, a previously healthy 3 year-old boy is noted to have a resp. rate of 40 breaths/min, a HR of 140 beats/min, hepatomegaly and a gallop rhythm. No heart murmurs are detected. Of the following, the MOST likely diagnosis is:

A. Acute rheumatic fever
B. Infective endocarditis
C. Myocarditis
D. Paroxysmal atrial tachycardia
E. Pericarditis
A 13 year old boy who has a bicuspid aortic valve and who received treatment for dental caries about 3 weeks ago now complains of lethargy, decreased energy, and reduced appetite. Findings on physical examination include low grade fever, splinter hemorrhages, splenomegaly and a new murmur consistent with aortic insufficiency.

Among the following, the BEST study to confirm the diagnosis in this patient would be:

A. blood culture  
B. chest radiograph  
C. complete blood count  
D. transesophageal echocardiogram  
E. erythrocyte sedimentation rate

A 14 year old boy complains of dull chest pain over the precordium. It began 4 days ago and occurs intermittently. It is not associated with activity, but it does increase when he is in a supine position and decreases when he is leaning forward. The frequency, duration, and the intensity of the pain has been increasing. Among the following, the MOST likely explanation for these findings is:

A. acute rheumatic fever  
B. arrhythmia  
C. costochondritis  
D. myocardial ischemia  
E. pericarditis

An 8 year old girl’s parents complain that she has been hyperactive and somewhat irritable for 2 weeks. She has jerky sudden movements of the shoulders and seems to have great difficulty sitting still. On physical examination the MOST likely additional finding in this child is:

A. icteric sclerae  
B. mitral regurgitation  
C. Osler nodes  
D. severe hypertension  
E. splenomegaly
KAWASAKI DISEASE

- Fever of 5 days duration, enlargement of lymph nodes, mucositis, non purulent conjunctivitis, rash
- Thrombocytosis and elevated ESR seen in 2nd week
- Coronary aneurysms are the most common cardiac manifestation and occur during week 2.
- Treatment is IVIG 2gm/kg as a single dose and high dose aspirin.
- Steroids occasionally needed for cases unresponsive to IVIG.

A 9 week old infant has had irritability and fever to 104 F for 8 days. Cultures of blood, urine and cerebrospinal fluid are negative. A coalescing red maculopapular rash has been present on the trunk and extremities since the second day of the illness. Red scleral conjunctiva without exudate are noted.

Of the following, the MOST likely complication to develop is:

A. aortic thrombosis  
B. cerebral infarction  
C. coronary artery aneurysms  
D. renal vein thrombosis  
E. splenic infarction

You are leading teaching rounds with the residents at the hospital. They present an 18-month-old boy who has had 6 days of a temperature to at least 102.3° F (39.1° C). He also has nonexudative conjunctivitis, a polymorphous rash, erythema of his lips, and swelling of his hands and feet. The residents ask you to comment on the use of echocardiography in this condition.

Of the following, the MOST accurate statement about echocardiography in this disease is that:

A) abnormal results at diagnosis suggest a poor outcome  
B) it should be performed only if C-reactive protein concentrations are elevated  
C) it should be performed to confirm the diagnosis  
D) normal results at diagnosis obviate the need to repeat the study  
E) the study may be useful in confirming atypical cases
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D. normal results at diagnosis obviate the need to repeat the study
E. the study may be useful in confirming atypical cases

4-year-old girl, hospitalized 6 weeks earlier with Kawasaki disease, continues recommended treatment to reduce the risk of coronary artery aneurysm. This patient is at increased risk for which of the following complications if she becomes infected with an influenza virus?

A. Reye syndrome
B. Toxic epidermal necrolysis
C. Pseudotumor cerebri
D. Autoimmune hepatitis
E. Drug reaction with eosinophilia and systemic symptoms (DRESS) syndrome

Good Luck!!!