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Herma Heart Institute

A Cardiologist's perspective for kids and families

Anoop K. Singh, MB BCh

Medical Director of Pediatric Cardiology

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Thank you to Camp Odayin!!!

Warning: This is a total hodge-podge ☺

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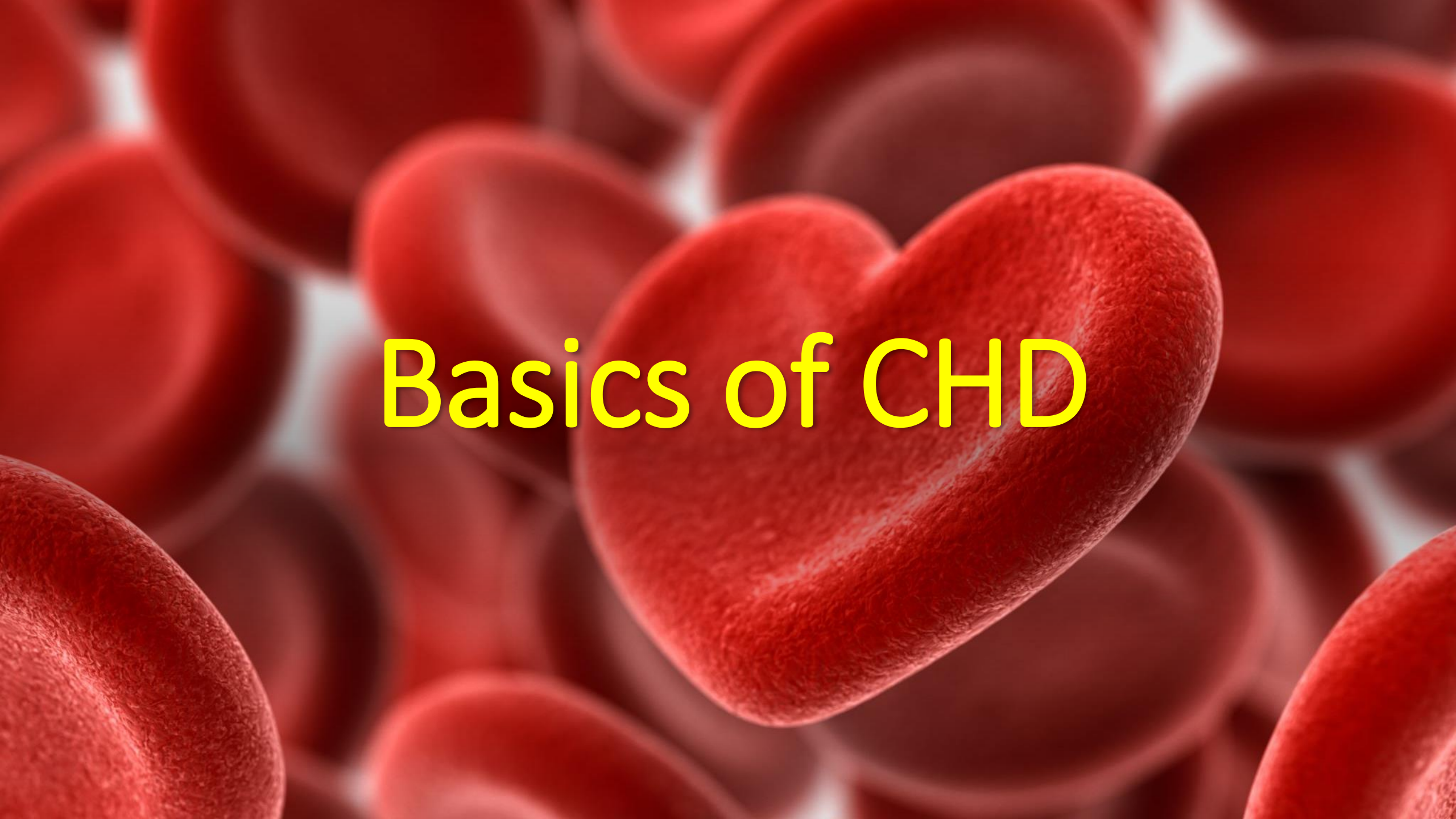
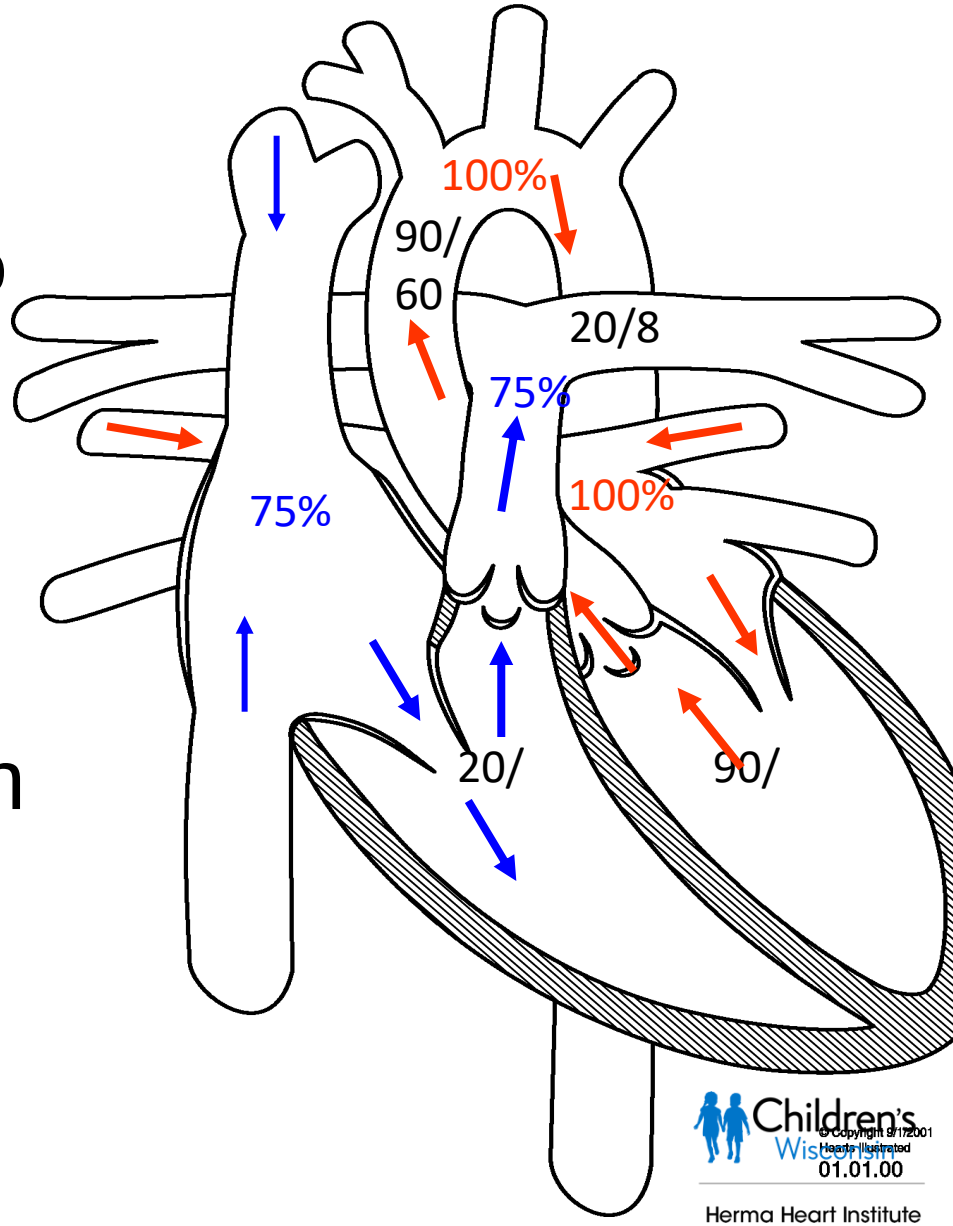
Abbreviations & Definitions

- CHD = congenital heart disease
- Echo = echocardiography (Ultrasound of heart)
- ECG = electrocardiogram
- Cath = cardiac catheterization (heart procedure typically using access through groin/nack blood vessels)
- Atria = smaller, “receiving chambers” in the heart; aka “top chambers”
- Ventricles = large, muscular pumping chambers in the heart; aka “bottom chambers”


Basics of CHD

How the heart works

- Right side pumps “blue blood” to the lungs
- Left side pumps “red blood” to the body
- 4 “one-way” valves keep blood flowing efficiently in one direction
- The heart’s electrical system coordinates all of this ☺



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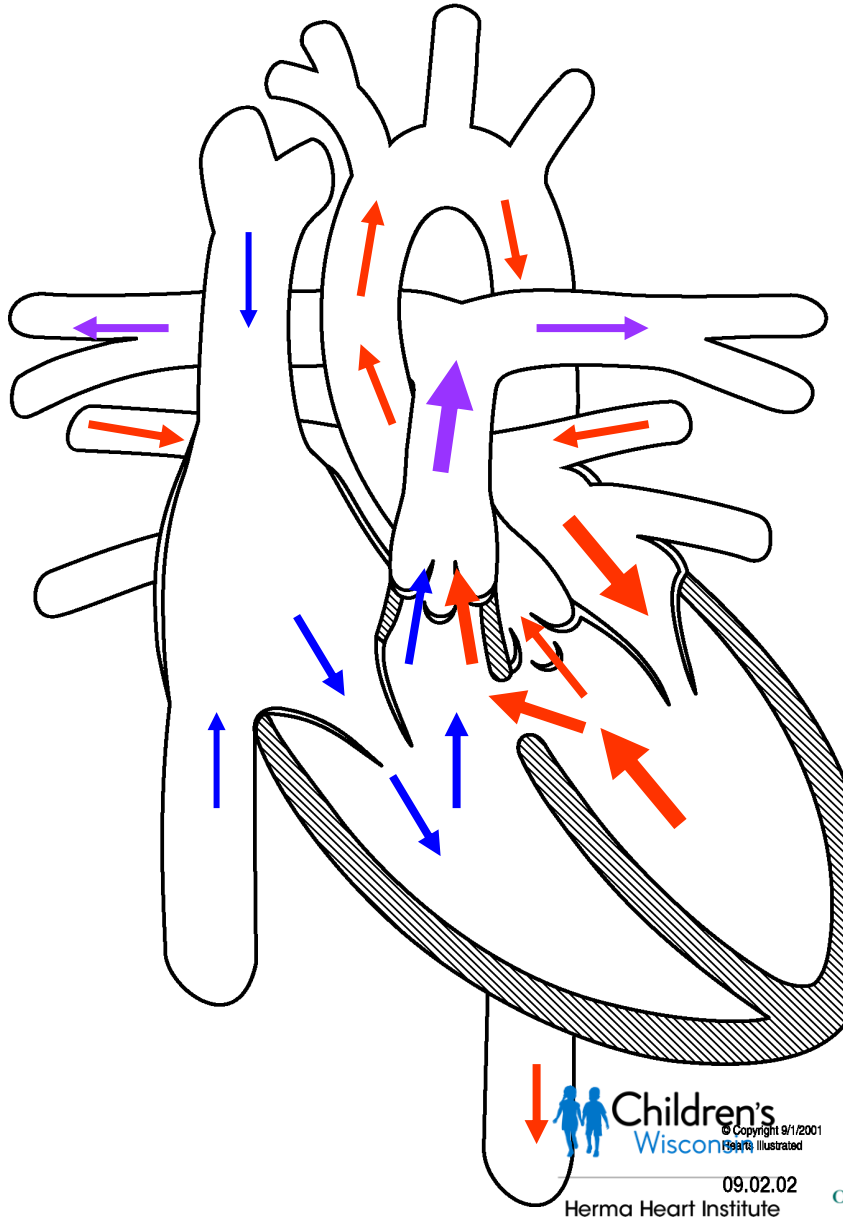
Basic CHD

A large hole results in:

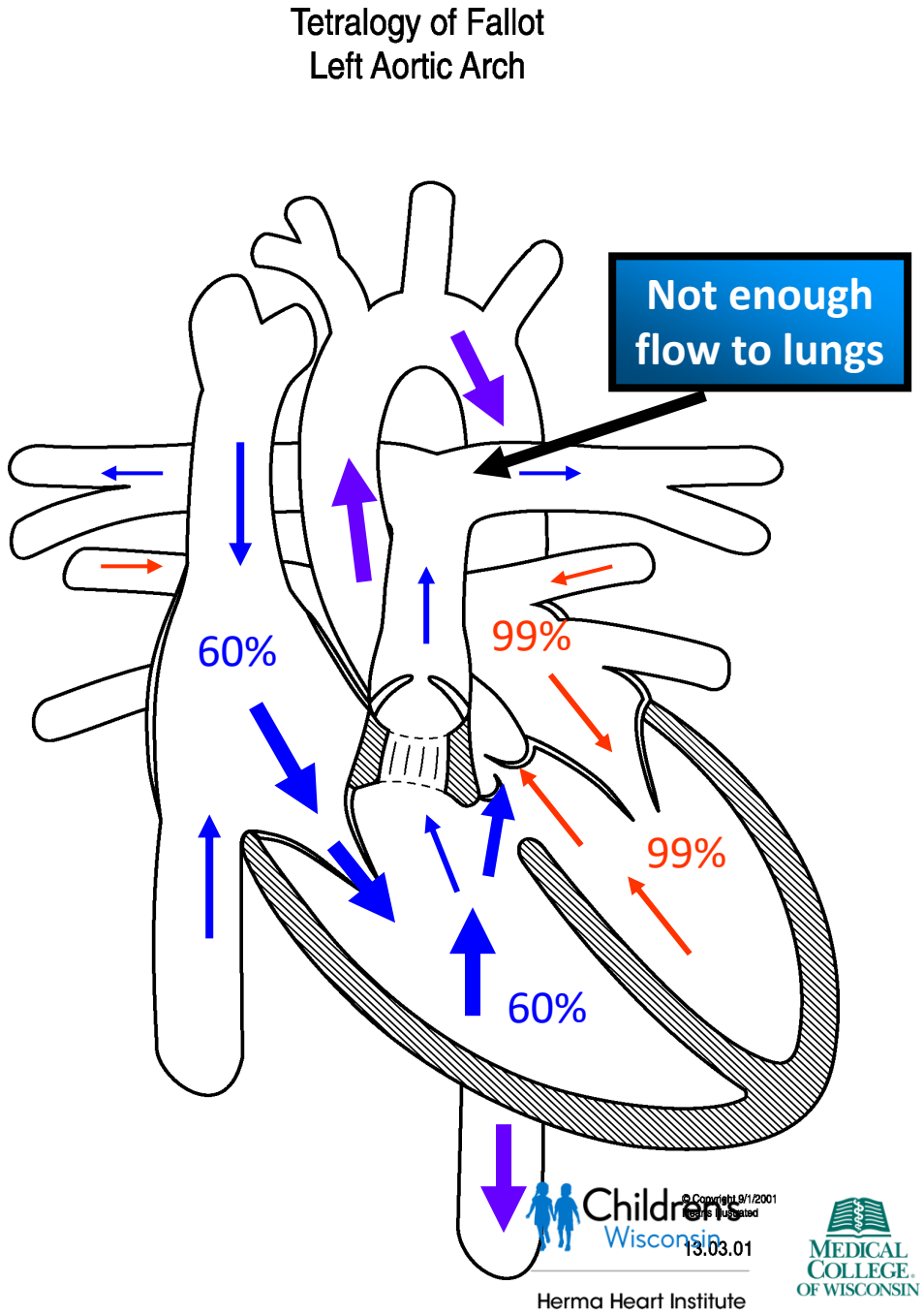
- Extra blood that inefficiently circulates to the lungs
- Increased work for the heart to compensate (always exercising)
- Challenges with feeding and growth



Moderate CHD

Large hole + obstruction to flow to the lungs:

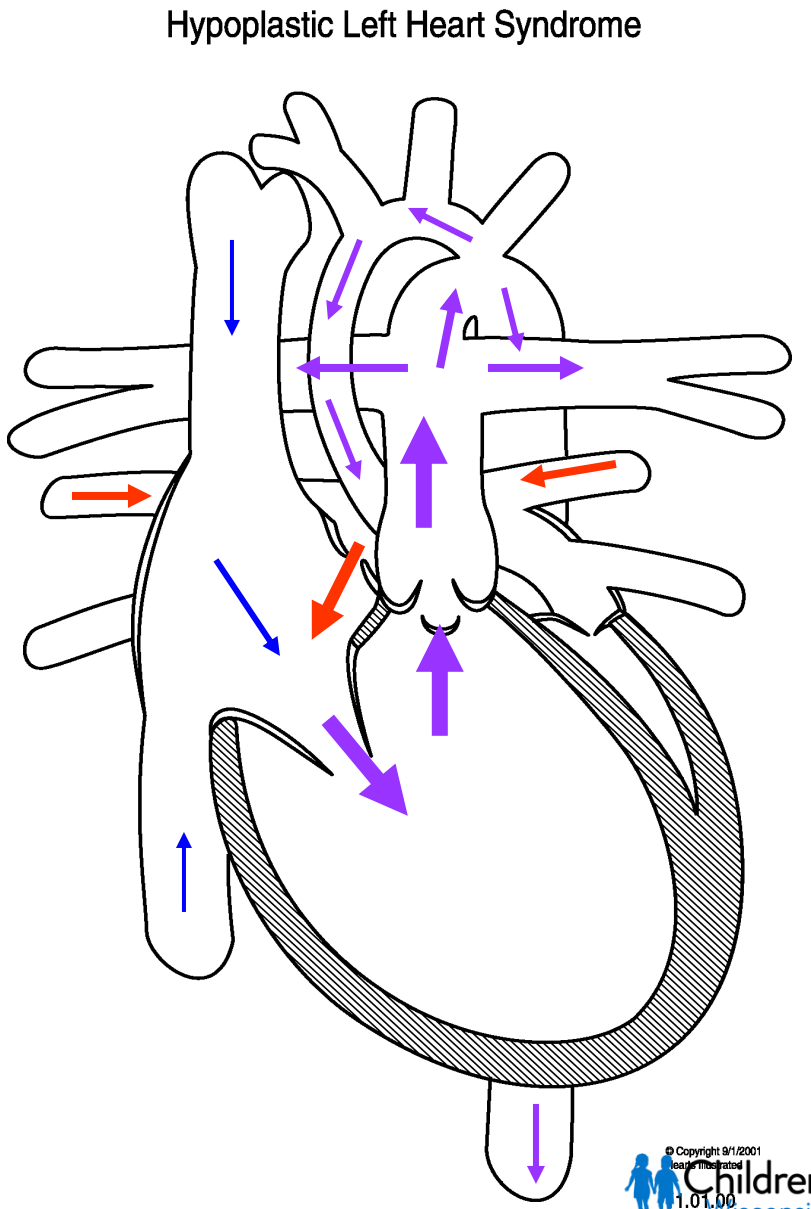
- Less issues with poor feeding
- But now less oxygen delivered to the body
- Risk of abrupt, life threatening episodes



Complex CHD

One ventricle has to pump to both the lungs and body:

- Increased strain on one ventricle
- No perfect ability to split blue from red blood
- Typically involves a series of surgeries over years to best optimize blood flow



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A fetal diagnosis of a heart problem

- Nowadays most CHD is diagnosed prenatally
- Many conditions are clear cut by the fetal study but all require an echo after birth to confirm and clarify some details
- A plan is often put in place for steps to take after the baby is born:
  - Place of delivery
  - The team that needs to be present
  - The need for critical medications
  - The need for critical procedures (rare)

A diagnosis of CHD after birth

Critical CHD

- Babies can be very sick on discovery of the CHD
- Stabilize at site of initial presentation and then transport to a CHD center

Not Critical CHD

- Often diagnosed in clinic

Various categories of CHD

1. Critical CHD that may involve a high-risk surgery but has the chance for fairly normal heart function
2. Critical CHD where only a cath intervention is needed
3. Critical CHD where the problem requires high-risk surgery and the heart problem cannot be completely “normalized”
4. Critical CHD where the surgery is not as high-risk
5. Complex CHD – single ventricle
6. Complex CHD – two ventricles (or biventricular)

Various categories of CHD (cont.)

7. Infant CHD typically requiring one surgery
8. Infant CHD with high likelihood of multiple surgery/intervention
9. Infant CHD that is monitored and may not require surgery
10. Childhood CHD that requires surgery or cath intervention
11. Childhood CHD that may not cause symptoms but carries risk for cardiac arrest

(And so many more that do not fit in any of these categories)

Management of CHD

Reasons a doctor recommends treatment

Symptoms

- A clear cut reason to treat – make the child feel better
- Net effect of treatment is benefits > risks
- Without treatment the condition could worsen

No Symptoms

- Although not causing problems now, it may be an issue if left unchecked
- Issue unlikely to resolve on its own
- May miss a window of safe opportunity to treat
- A risk for sudden, unexpected events

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Questions parents should consider:

- What is the risk-benefit profile?
- (*If a surgery or cath intervention*) Is there data on patient outcomes at your center?
- Is this a short-term, long-term, or lifelong treatment?
- How clear cut is the decision? “95% certain” or more of a “not sure but we think this is the best option”?
- Is this what most cardiac providers would recommend?
- (*If not a clear cut decision*) Should I seek a 2nd opinion?

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Treating Before Symptoms

- Often accepting some risk up front for long term benefit
- There may be a “missed window” if waiting too long
- Compliance is a big issue as the child/teen sees no benefit
- What if we are wrong about the risk profile?
- Overall, a lack of data in pediatric cardiology

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DATA

1. CHD Uniqueness

- While children with the same CHD can be grouped together, there is still some variation in the specific anatomy (eg- Tetralogy of Fallot (ToF):
  - “Standard ToF”
  - “Pink ToF”
  - ToF with pulmonary atresia
  - ToF with absent pulmonary valve
  - ToF + AV canal defect
- As such any centers surgical results on ToF can vary depending on which subtypes they include in their data

2. Non-CHD Problems

- Many children with CHD have additional diagnoses that can have minor or major impact on the prognosis and outcomes:
  - Genetic diagnoses
  - Impact of other organ systems problems

3. Surgeons are unique "variables"

- Pediatric cancer care can be very data-driven as many patients are enrolled in clinical trials
  - Variables are more controlled (eg- drugs, radiation)
- While cardiac surgeries may have the same name for the same type of CHD often there are nuances in each case for the surgeon
- Even within the same cardiac center, different surgeons have different approaches to the same patient

Despite all this we have some data

- A lot of research focuses on understanding the best management for CHD
- In addition to research there is quality improvement (QI) work which looks to standardize processes as much as possible
- Cardiac centers collaborate to look at how their center's outcomes compare to other national heart programs
- The "ultimate" would be Precision Medicine which would aim to provide unique treatment plans for children based on their genetics, lifestyle, etc

Making decisions with not much data

- It's hard (but not nearly as hard as what the patient and family are going through).
- Is it just me or does no one have the answer to this problem?
- Can I extrapolate from available data or my past experiences?
- Will this family trust me since I just answered "I'm not sure" multiple times in this conversation?
- At the same time giving up is not an option so coming up with a plan is important (even if that plan is "doing nothing").

Longitudinal Care for CHD

Hospital-based Clinics

- All testing available: echo, ECG, stress, labs
- Ability to do sedated echos
- Can see multiple providers at once, both within and outside cardiology
- If needed, the ability to admit to the hospital, send to the ER
- Association for the child and family with their surgeries, hospitalizations, etc
- Parking, traffic, etc
- Longer trip for many
- Hospital-based facility fees

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Non-hospital-based Clinics

- Often easier to navigate
- Closer to home for many
- Should all have echo and ECG capability

- Less ability to see multiple providers
- Testing is typically more limited (likely no sedated echo)

Echo

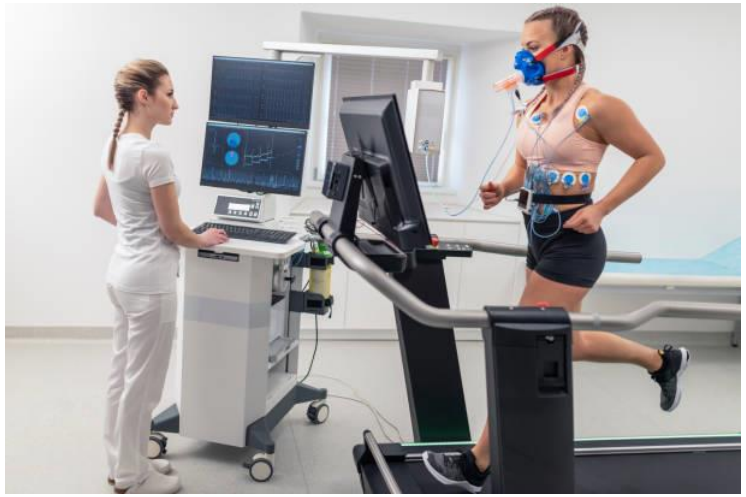
- The most important test for CHD
- Provides anatomy but also a very functional assessment of valve function, heart pump strength, and changes in heart size
- Can take 15-60 minutes depending on condition and situation
- Babies cannot lie still for that duration so sometimes sedation is used to get high quality information
- Ultrasound requires “good acoustic windows” so sometimes, (and not anyone’s fault) good pictures cannot be obtained

Cardiac MRI & CT

- Cardiac MRI can be a 1-2 hour study – no radiation
- Cardiac CT < 15 minutes – radiation
- For kids < 8 yo typically need sedation/anesthesia
- Very high quality, anatomic and functional studies that have replaced a lot of diagnostic caths for kids with CHD

Exercise Stress Tests

- Typically either a treadmill or stationary bicycle is used
- An ECG is hooked up and regular blood pressure checked
- Can connect a mouthpiece to measures normal breathing
- Combined the test can:
  - Measure the capacity for heart and lung function
  - Assess or unmask symptoms related to exercise



Thoughts on various follow-up scenarios

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No surgery as infant but might need it later (eg- small hole in the heart)

- Implication is that the CHD is unlikely to cause much symptoms but may be a problem later
- Most of the time the decision to proceed with surgery or cath will be based on the echo or MRI (and not symptoms)
- Sometimes there are “vague symptoms” (ie- we cannot discern if it’s related to the CHD or not) and that may or may not decide the timing for intervention
- Often this follow up is at 6 or 12 month intervals

No surgery as infant but will have future intervention

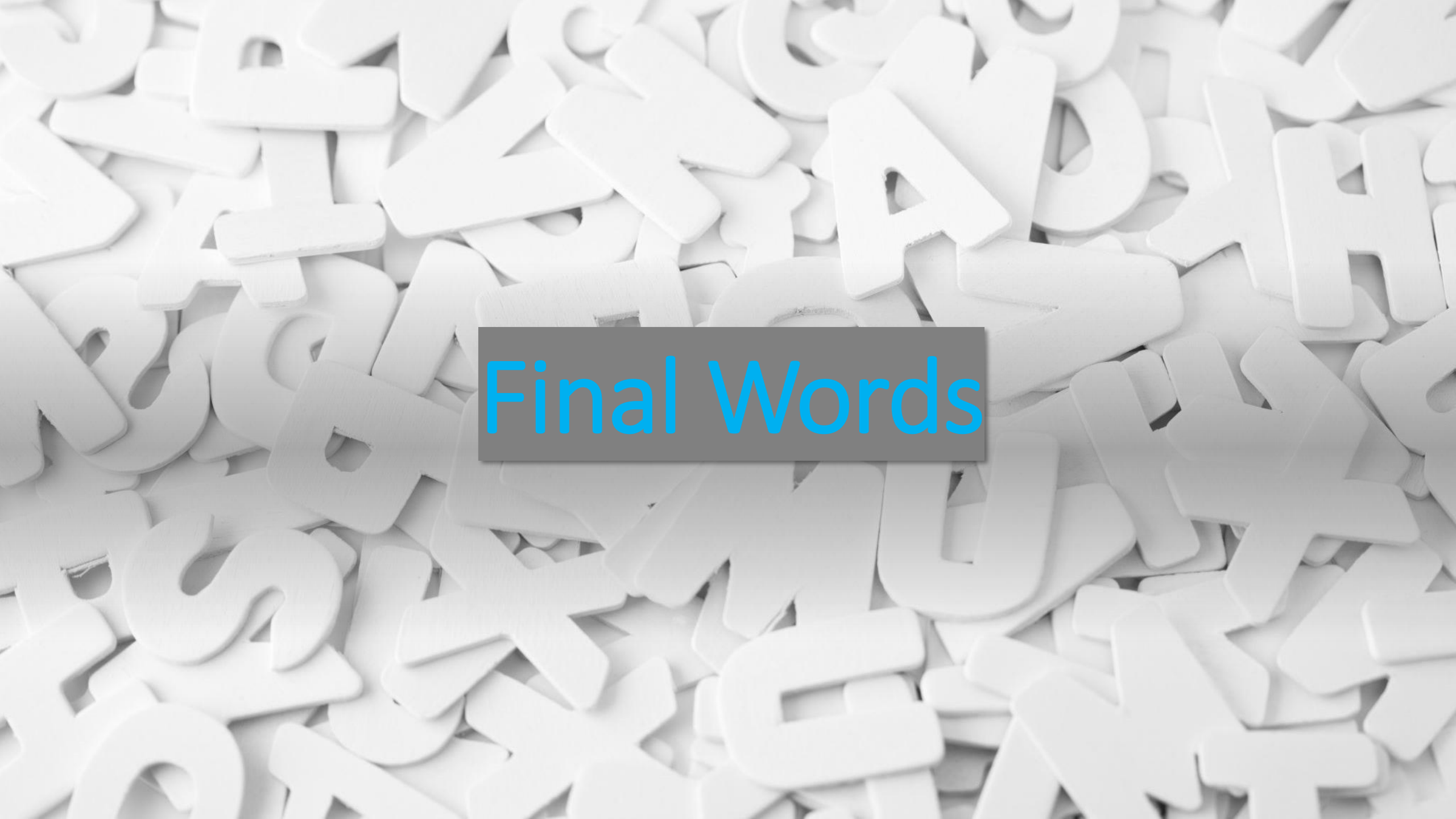
- Mild types of CHD (eg- isolated ASD)
- Why not do intervention earlier ("just fix the heart")?
  - A larger patient decreases need for blood product exposure
  - Larger patient = larger heart = easier for surgeon
  - Brain is more fully developed if heart-lung bypass surgery is needed
  - Once bigger, cath options may be available
- Why not do intervention later?
  - Changes in heart (eg- enlargement) may not reverse despite surgery
  - New problems (eg- arrhythmias) may develop
  - Some ages (3-6 yo) are best developmentally for hospitalization

Had surgery and will need future surgery (eg- artificial valve)

- While the child and their heart grows, the artificial valve will not
- Both symptoms and imaging often play a role in deciding on the timing for next intervention
- Deferring intervention may afford option to place largest replacement valve vs risk of waiting too long

Adult care for CHD

- Cardiologists that care for adults deal with many issues but CHD is a rare one
- Specialized cardiologists are trained in the care of both CHD and adult medicine – ACHD Specialists
- How do you find one?
  - Some have this skillset, regardless of board qualifications, based on practical experience over many years
  - Most ACHD providers now will have done additional training in the field and be board certified (American Board of Internal Medicine website)


Final Words

What to look for in CHD care

- How easy is it to communicate with the CHD team?
  - Generic operators with multiple transfers or a dedicated number for cardiac patients?
  - Who picks up the phone?
  - Do they have a non-calling system (eg- Epic's MyChart)?
- Are testing and the doctor's visit easily coupled?
- Does the doctor know when to seek advice from colleagues?
- Do they listen to you? Are they good with your child?

TRUST

If surgery or cath intervention needed

- Does this center do a lot of this procedure?
- Meet with the surgeon or interventionalist (my recommendation)
- Post-op care should be driven by the ICU, surgeon, and cardiologists (in that order, IMO)
- How do they prepare you for the transition to care at home?
- Is your child's cardiologist connected and apprised of what is going on throughout the hospital stay? (important for that first follow up clinic visit)

What you can do

- Try and figure out the capabilities of the ER closest to your home (especially if your CHD center is > 45 minutes away)
- Know infant, child, adult CPR – a skill you never want to use
- Work with your school to help them understand your child's condition (and have your CHD team help you with that)
- Ask your school if they are prepared for cardiac emergencies (eg- Project ADAM HeartSafe Schools)