

Cardiovascular Medicine in the Cascades

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**WHAT HAPPENS TO KIDS WITH
CONGENITAL HEART DISEASE:
A GROWING PROBLEM**

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Outline

1. Demographic of congenital heart disease.
2. Classification of congenital heart disease into risk groups.
3. Common issues in the adult with CHD.
4. Summary.

Incidence of Congenital Heart Disease in USA and Oregon

	USA	Oregon
Population	300 million	3.7 million *
Live Births/Year	4.2 million	46,250 (12.5 births/1000 population)
Births/Year with CHD	50,000 **	555 **

* July 2006.

** Incidence of 1.2% (1 per 84 births)

Prevalence of Congenital Heart Disease in USA and Oregon

	USA	Oregon
Total number with CHD	1.33 million	16,000
Adults (>18 years old) with CHD	838,000	10,000

There are now more adults with congenital heart disease than children.

Prevalence of Congenital Heart Disease in Deschutes County

- Deschutes County population in 2006 was 130,000 (4.0% of Oregon population).

Prevalence of Congenital Heart Disease in Deschutes County

	USA	Oregon	<i>Deschutes County</i>
Total number with CHD	1.33 million	16,000	575
Adults (>18 years old) with CHD	838,000	10,000	380

Prevalence of Coronary Artery Disease in USA and Oregon

	USA (Pop = 300 M)	Oregon (Pop = 3.7 M)
Stable Angina	6.4 million	79,000 thousand
Acute Coronary Syndrome	7.1 million	88,000 thousand
TOTAL	13.5 million	167,000 thousand

Prevalence of Congenital Heart Disease vs Coronary Artery Disease in USA and Oregon

	USA	Oregon
CHD	1.33 million	16,000 thousand
CAD	13 million	167,000 thousand

Congenital Heart Disease vs Coronary artery Disease in USA and Oregon

	Congenital Heart Disease	Coronary Artery Disease
Age at diagnosis	Infancy	Men = 62 years Women = 70 years
Duration of care	40-60 years	15 years

Congenital Heart Disease vs Coronary Artery Disease in USA and Oregon

	Congenital Heart Disease	Coronary Artery Disease
Age at diagnosis	Infancy	Men = 62 years Women = 70 years
Duration of care	40-60 years	15 years
<i>Patient years of care provided</i>	<i>66.5 million</i>	<i>195 million</i>

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Low Complexity ACHD

(Very good survival and few complications)

- Patent ductus arteriosus (PDA)
- Atrial septal defect (ASD), small or moderate size
- Pulmonary valve stenosis (PS), mild
- Ventricular septal defect (VSD)
- Aortic stenosis (AS), mild

Moderate Complexity ACHD

(Good survival; moderate risk of complications)

- Large ASD in adult
- Pulmonic stenosis (PS) \geq moderate
- Coarctation of the aorta
- Aortic stenosis (AS) \geq moderate
- AV septal defect (aka endocardial cushion defect, AV canal)

High Complexity ACHD

(Reduced survival and high risk of complications)

- Cyanotic lesions, repaired
 - Transposition of the great arteries (TGA) (1,2)
 - Congenitally corrected TGA (1,2)
 - Truncus Arteriosus (2)
 - Tricuspid Atresia; Mitral Atresia (1,2,3)
 - Pulmonary Atresia (2,3)
 - Double outlet right ventricle (DORV) (2)
 - Common ventricle anatomy (2,3)
- Eisenmenger Syndrome (2)
 - (1) Systemic right ventricle
 - (2) Conduit and/or valve placed as part of repair
 - (3) Fontan surgery (2 ventricle repair not possible)

Congenital Heart Disease in USA

Lesion Severity	≤ 15 Years of age	> 15 Years of age	Totals
Mild	200,000	750,000	950,000 (58%)
Moderate	120,000	360,000	480,000 (29%)
Severe	110,000	100,000	210,000 (13%)
Totals	415,000	1,210,000	1,640,000

Survival Data

- *Am J Cardiol* 2000;86:1111-1116
2,609 patients followed at the University of Toronto Congenital Cardiac Center for Adults over 15 years (1981-96)

Survival-Low Complexity CHD

15 Year Survival

PDA	96%
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ASD	93%
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PS	99%
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VSD	93%
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Survival-Moderate Complexity CHD

	<u>15 Year Survival</u>
COA	98%
AS	97%
AV Septal	88%

Survival-High Complexity CHD

15 Year Survival

Tetralogy	93%
d-TGA	90%
Other Cyanotic Lesions	77%

Causes of Death in ACHD

- Cardiovascular Death (83%)
- Non-cardiovascular Death (17%)

Am J Cardiol 2000;86:1111-1116

Causes of Death in ACHD

- Sudden 26%
- CHF 21%
- Peri-operative 18%
- Other 18%
 - Pul emboli
 - Bleeding
 - MI

Value of Categories in ACHD

- Incidence of medical problems is related to complexity of underlying CHD
- Identifies individuals that need close follow-up

Frequency of Follow up in Adults with CHD

- Low complexity ACHD
 - Infrequent visits (3-5 years) to primary care physician, internist, or cardiologist
- Moderate complexity ACHD
 - Annual ACHD clinic visits
- High complexity ACHD
 - Close follow-up in ACHD clinic (6 months)

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Ongoing Problems In ACHD

- Arrhythmias
- CHF
- Pregnancy
- Cardiac surgery/intervention
- Endocarditis
- Non-cardiac surgery
- Activity, job, insurance, disability

Arrhythmias

- Most common reason to see M.D.
- Increase with age & complexity of CHD
- Presentation may be subtle (not feeling well, tachyarrhythmia that is slower than usual SVT, CHF)

Arrhythmias

- Supraventricular arrhythmias much more common than ventricular
- Wide QRS tachyarrhythmia is usually supraventricular. (EPS may be required to distinguish SVT with wide QRS from ventricular tachycardia)

Arrhythmias

Arrhythmias are
commonly
precipitated by
hemodynamic
problem

Arrhythmias

- Evaluate for and correct hemodynamic abnormality
- Restore NSR
- Prevent future arrhythmias
(antiarrhythmic agents, catheter ablation, AICD)
- Anticoagulation issues

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CHF

CHF commonly
precipitated by
arrhythmias or
hemodynamic
problems

CHF

- Correct arrhythmias and hemodynamic abnormality.
- Usual medical therapy for LV dysfunction.
- Systemic RV dysfunction treated same as LV, but not as effective.
- Heart transplantation is an option. 5 year survival in CHD is 64%, only slightly worse than CAD (69%) or cardiomyopathy (71%).

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Pregnancy

- Underlying heart disease is the second leading cause of death in pregnancy (suicide first)
- Large number of women with CHD in reproductive years
- Discuss reproductive issues including contraception
- Need to provide advice about the risk of pregnancy

Maternal Death during Pregnancy

OHSU

Country	Deaths/100,000 confinements	Death/number of confinements
Sweden (best)	2.0	1/50,000
Australia	8.2	1/12,200
UK	11.4	1/8770
USA	17.0	1/5880
Global Average	400	1/250
Women + Heart Disease	500	1/200
Sierra Leone (worst)	2,000	1/50

Pregnancy

Three common questions

- What is the risk my child will have congenital heart disease?
- Can I safely have a child?
- Can I have a vaginal delivery?

Pregnancy

- Mendelian inheritance (50% chance of disease)
 - Marfan Syndrome
 - Noonan Syndrome (ASD and Dysplastic PV)
- Non-Mendelian inheritance
 - Incidence of births in USA with CHD is 1%
 - If mother has CHD, risk in offspring is 5-7%; lower if father has CHD with risk at 2-4%

Pregnancy

- Identify women with cardiac abnormalities associated with very high mortality risk (7-50% vs average mortality of 0.01% in developed countries).
- These women are advised to
 - avoid pregnancy
 - fix abnormality if possible prior to pregnancy
 - consider early termination if pregnant (women rarely chose to do this)

Pregnancy

High Mortality Risk Group

1. Pulmonary hypertension (RVsp >70 mmHg or >3/4 systemic pressure)
2. Hypoxemia (oxygen saturation <85%)
3. Eisenmenger syndrome
4. Severe systemic ventricular dysfunction (EF <35%); NYHA class III or IV; Max $\dot{V}O_2$ <20 ml/kg/min)

Pregnancy High Mortality Risk Group

5. Aortic root >40 mm diameter in Marfans or other aortopathies.

6. Severe left heart obstruction
 - Aortic stenosis (AVA <1.0 cm², mean gradient >50 mmHg)
 - Mitral stenosis (MVA $<1.2-1.5$ cm², mean gradient >10 mmHg)
 - Coarctation (gradient >30 mmHg), supra- or sub-valvular obstruction)

Pregnancy

- In remaining women, provide estimate of risk for pregnancy after thorough evaluation
- Recommend method from Siu et al (Circ 2001;104:515-521)
 - Validated
 - Not lesion specific

Pregnancy

- In almost all cases, vaginal delivery is an option (equivalent mortality, but lower morbidity vs C-section)
- Cesarean section is best done for
 - Obstetrical indication
 - Specific cardiac indications
 - Prolonged INR (on warfarin) at time of delivery
 - Pulmonary hypertension
 - Enlarged or unstable aorta
 - Severe obstructive left heart lesions
 - Heart failure with limited cardiac reserve

Pregnancy

Risk of C-section vs Vaginal Delivery

- Increased thromboembolic events (3 fold)
- Increased risk of infection/sepsis (10 fold)
- More blood loss (CS = 500-800cc vs VD = 300-500 cc) and greater need for transfusion (CS = 5.2% vs VD = 2.1%)
- Longer hospital stay (CS = 5 days vs VD = 1-2 days)

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Additional Cardiac Surgery or Interventions

- Reoperation needed for
 - obstruction
 - degeneration of valves (native/prosthetic) or conduits
 - Damage due to endocarditis
- Team approach/experience essential

Cardiac Surgery/Intervention

- Mortality and morbidity increase with multiple operations.
- Re-opening the chest associated with significant bleeding.
- Scar and distorted anatomy make finding a desired anatomic site a significant challenge.

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Endocarditis

- THINK OF ENDOCARDITIS (20-35% of IE cases occur in CHD patients). Problems are
 - Delayed diagnosis (not thinking of IE)
 - Antibiotics prior to obtaining blood cultures
- Risk of endocarditis is higher in adults than children with CHD
- Major portal of entry is still oral cavity
- Regular flossing, brushing, and dental care essential to reduce risk

New Guidelines for Endocarditis Prophylaxis*

- Mechanical and bioprosthetic heart valves
- Previous endocarditis
- Cardiac transplantation recipients who develop cardiac valvulopathy.

* *Circ* 2007;115:1-19

New Guidelines for Endocarditis Prophylaxis*

- Congenital heart disease with
 - Unrepaired cyanotic heart disease including palliative shunts and conduits
 - Repaired with prosthetic material or device (surgery or by catheter intervention) during the first 6 months after the procedure
 - Repaired with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device

* *Circ* 2007;115:1-19

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Non-cardiac Surgery

- Team approach/experience with CHD minimizes risks
- Concerns during peri-op period
 - Air emboli from IV when shunt present
 - Fluid management
 - Bleeding abnormalities
 - Infection/Endocarditis
 - Arrhythmias
 - Death

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Special Issues

- **Activities:** provide guidance
- **Jobs:** match job with physical capacity, letters of support to employers (still job discrimination)
- **Insurance:** Health and life insurance rates are increased in almost all CHD cases and neither may be available to those with high risk CHD
- **Disability:** support when indicated

OUTLINE

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Summary

- Most children with congenital heart disease survive into adulthood. More adults with CHD are alive than children with CHD.
- This “growing” problem has led to a new subspeciality in cardiology of adult congenital heart disease.
- Their medical problem is life-long.
- The more complicated lesions have a high potential for major problems due to their congenital heart disease. Few will be without problems.

Thank you