CONGENITAL HEART DISEASE

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Embryology

Cardiogenic Region ~16 days

Sulik KK, Bream PR. Sulik collection, UNC
www.med.unc.edu/embryo_images
Heart Begins Beating ~22 days

Sulik KK, Bream PR. Sulik collection, UNC
www.med.unc.edu/embryo_images
Embryology

Straight Heart Tube ~24 days

Sulik KK, Bream PR. Sulik collection, UNC
www.med.unc.edu/embryo_images
ATRIAL SEPTAL DEFECT
ATRIAL SEPTAL DEFECT

- 4\textsuperscript{th}-6\textsuperscript{th} WEEK OF GESTATION, THE SINGLE ATRIAL CHAMBER IS DIVIDED INTO TWO

- SPACE BETWEEN THE TWO SEPTUM IS OSTIUM PRIMUM, OR FIRST HOLE.

- FENESTRATIONS APPEAR IN CENTER LEADING TO SECOND HOLE- OSTIUM SECUNDUM.
ATRIAL SEPTAL DEFECT

- ULTIMATE BALANCE BETWEEN PROLIFERATION AND ABSORPTION OF THE TWO SEPTA –FORAMEN OVALE.
ATRIAL SEPTAL DEFECT

Wall between the atria doesn't close completely leading to a mixture of oxygenated and de-oxygenated blood.
4 Left-to-Right Shunts

A

OSTIUM PRIMUM

SEPTUM SECUNDUM

ENDOCARDIAL CUSHION

INTERVENTRICULAR FORAMEN

B

OSTIUM SECUNDUM

SEPTUM PRIMUM

INTERVENTRICULAR FORAMEN

C

SEPTUM SECUNDUM

FORAMEN OVALE

INTERVENTRICULAR FORAMEN

D

SEPTUM PRIMUM

INTERVENTRICULAR SEPTUM MUSCULAR

INTERVENTRICULAR SEPTUM MEMBRANOUS
ATRIAL SEPTAL DEFECT

• SINUS VENOSUS DEFECT
• CHIARI NETWORK
• OSTIUM SECUNDUM
• OSTIUM PRIMUM
ATRIAL SEPTAL DEFECT

• ERROR IN DEVELOPMENT IN DEPOSITION OR ABSORPTION, A COMMUNICATION RESULTS-ASD.
• IF HIGH IN SEPTUM NEAR SVC AND IF RT PULM. VEIN IS ANOMALOUS-Sinus Venosus Defect.
ASD-SECUNDUM

• MOST COMMON TYPE OF ASD
• DEFECT IS LOCATED IN CENTER OF SEPTUM.
ASD-PRIMUM

• COMMUNICATION IS AT THE LOCATION OF THE LOWER END OF SEPTUM.

• USUALLY ASSOCIATED WITH DEFECT IN THE MITRAL VALVE.

• CLASSIFIED AS INCOMPLETE AV CANAL OR PARTIAL ENDOCARDIAL CUSHION DEFECT.
HEMODYNAMICS
ASD

- LEFT TO RIGHT SHUNT
- INCREASED RIGHT SIDED VOLUME
- RESULTS IN DILITATION OF RA, RV AND PULMONARY VESSELS
- LEFT HEART IS UNCHANGED!
PHYSICAL EXAM

• EJECTION MURMUR-2\textsuperscript{ND} LEFT INTERCOSTAL SPACE

• SAME VOLUME OF BLOOD!!!

• MID-DIASTOLIC FILLING WITH FIXED VOLUME CONSISTENTLY DELAYS CLOSURE OF PULMONIC VALVE

• FIXED SPLIT SECOND SOUND
PHYSICAL EXAM

• OSTIUM PRIMUM DEFECT
• SAME DEFECTS AS SECUNDUM WITH ADDITION OF MITRAL REGURGITATION.
• POOR GROWTH, CHF-INFANT
VENTRICULAR SEPTAL DEFECT
VSD

• DEVELOPES BETWEEN THE 4TH AND 8TH WEEKS OF GESTATION

• SINGLE VENTRICLE IS DIVIDED IN TWO.

• TWO PORTIONS OF SEPTUM-MEMBRANOUS AND MUSCULAR
HEMODYNAMICS OF VSD
VSD

- LEFT TO RIGHT SHUNT
- LV TO RV TO PULMONARY ARTERY
- RA-NORMAL IN SIZE
- RV DILATES AS DOES MAIN PA, LEFT ATRIUM AND LEFT VENTRICLE
VSD-PHYSICAL EXAM

• ONSET OF SYSTOLE PRODUCES HOLOSYSTOLIC MURMUR
• HEARD BEST AT THE 4TH LEFT ICS
• WIDESPREAD TRANSMISSION EVEN INTO PULMONARY ARTERY.
• LOUD!!!
• RV HEAVE
EISENMENGERS COMPLEX
EISENMENGER’S COMPLEX

• CLINICAL SITUATION WHERE IN A PATIENT WITH ANY LEFT TO RIGHT SHUNT DEVELOPS SUFFICIENT PULM VASCULAR DISEASE AND PHTN TO PRODUCE REVERSAL OF FLOW AND THEREFORE A RIGHT TO LEFT SHUNT
HEMODYNAMICS

- EQUILIZATION OF PRESSURES
- MURMUR DIMINISHES DUE TO LESS SHUNTING
- RV EJECTION TIME DIMINISHES PERMITTING THE PULMOSARY VALVE TO CLOSE SOONER AND INTENSITY OF S2 INCREASES
HEMO’S (CON’T)

• WITH TIME, PULMONARY RESISTANCE INCREASES AND EXCEEDS SYSTEMIC RESISTANCE AND THE SHUNT THROUGH THE VENTRICULES REVERSES RESULTING IN SYSTEMIC CYANOSIS
PATENT DUCTUS ARTERIOSUS
PDA

• DEVELOPES BETWEEN THE 5TH AND 7TH WEEKS OF GESTATION

• AORTIC ARCH DEVELOPS WITH PROLIFERATION FROM APEX OF TRUNCUS ARTERIOSUS.

• ON THE LEFT, THE DISTAL PORTION MAINTAINS ATTACHMENT TO AORTA AND BECOMES DUCTUS ARTERIOSUS
ANATOMY OF PDA

• IN FETAL LIFE, DUCTUS SERVES AS A FUNCTIONING CONNECTION BETWEEN THE PULM ARTERY AND AORTA.

• AFTER BIRTH, THE PARTIAL PRESSURE OF O2 RISES AND THE PULM ARTERIOLES DILATE CAUSING THE DUCTUS TO CLOSE.
ANATOMY (CON’T)

• ULTIMATELY, THE DUCTUS FIBROSES AND BECOMES THE LIGAMENTUM ARTERIOSUM

• WHEN IT DOESN’T CLOSE IT IS CALLED A PATENT DUCTUS ARTERIOSUS(RE DUNDANT)
HEMODYNAMICS OF PDA
HEMODYNAMICS OF PDA

• HIGH PRESSURE AORTA COMMUNICATES WITH LOW PRESSURE PULMONARY ARTERY
• INCREASES VOLUME IN LUNGS AND SUBSEQUENTLY INTO LV
• SIMILAR TO VSD
HEMODYNAMICS

• RA, RV NO CHANGE
• MAIN PA, PULM VESSELS, LA, LV AND AORTA DILATE
CLINICAL APPLICATION

• OCCURS EARLY IN LIFE

• INITIALLY MURMUR IS SYSTOLIC, BUT AS DIASTOLIC EQUILIBRATION OCCURS, MURMUR BECOMES A CLASSIC TO AND FRO OR CONTINUOUS MURMUR OCCURS.
PHYSICAL EXAM OF PDA

• CONTINUOUS MURMUR
• LEFT OF STERNUM AT 2\textsuperscript{ND} OR 3\textsuperscript{RD} INTERSPACE
• COURSES ALONG STERNUM AND ALONG PULMONARY ARTERY
• DISPLACE APEX DUE TO INCREASED VOLUME WITH A THRUST
CLINICAL APPLICATION

• BIRTH-NORMAL
• 2-6 WEEKS-FLOW BEGINS AND SYSTOLIC MURMUR IS HEARD
• AS LA AND LV SIZE INCREASE, CHF DEVELOPES
• MURMUR BECOMES CONTINUOUS AS PA AND LV DIASTOLIC PRESSURES EQUAL
COARCTATION OF THE AORTA
COARCTATION

- 5th and 7th weeks of gestation, the aortic arch develops
- At area of patent ductus, aorta develops improperly, leaving a restricted lumen.
- Location: proximal, at, or distal to insertion of ductus.
COARCTATION

• MORE COMMON IN MALES
• RIB NOTCHING OCCURS DUE TO PHYSICAL ERROSION OF THE UNDERSURFACE OF THE RIBS AS A RESULT OF INTERCOSTAL COLLATERAL CIRCULATION
• ASSOCIATED WITH BICUSPID AORTIC VALVE
TETRALOGY OF FALLOT
TETRALOGY OF FALLOT

• 3\textsuperscript{rd} TO 4\textsuperscript{th} WEEK, THE COMMON TRUNK DIVIDES INTO THE PULMONARY ARTERY AND THE AORTA.

• 4\textsuperscript{th} AND 8\textsuperscript{th} WEEK, THE VENTRICLE DIVIDES INTO TWO
TETRALOGY

• DEFINED BY FOUR FINDINGS
• 1) INFUNDIBULAR STENOSIS
• 2) VENTRICULAR SEPTAL DEFECT
• 3) RIGHT VENTRICULAR HYPERTROPHY
• 4) OVERRIDING OF THE AORTA
HEMODYNAMICS

• DIMINISHED BLOOD FLOW TO THE LUNGS AND INCREASED BLOOD FLOW TO THE BODY.

• DUE TO THE STENOSIS OF THE INFUNDIBULUM, PULMONARY FLOW IS DIMINISHED. THE OVERRIDING AORTA ACCEPTS MOST OF THE RV BLOOD.
CLINICAL APPLICATION

• THIS PRODUCES A RIGHT TO LEFT SHUNT AND THEREFORE PRODUCES CYANOSIS OF PERIPHERY.

• CHILDREN PRESENT WITH CYANOTIC HANDS AND FEET

• CHILDREN SQUAT TO ENHANCE FLOW BACK TO HEART TO OXYGENATE.