Surgical challenges in atrio-ventricular septal defect in grown-up congenital heart disease

Anatomy of Atrioventricular Septal Defect (AVSD)

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Disclosures: None

Website: www.rbht.nhs.uk/cardiacMorphology
Anatomy of Atrioventricular Septal Defect (AVSD)

AV canal; endocardial cushion defect; ostium primum ASD

- 3% of all congenital heart defects
- Strong association with Down syndrome: ~ 30%
  - complete AVSD: 70-80% with Down
  - partial (primum) AVSD: 10% with Down

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Anatomy of AVSD

The atrioventricular junction

Normal heart (figure of 8)

AVSD

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Anatomy of AVSD

Faletra F, Nucifora G, Ho SY. Circ Cardiovasc Imaging 2011

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Anatomy of AVSD - biventricular AV connections

4 chamber view

Normal
Off-set between tricuspid and mitral valves

AVSD
Loss of off-set between right and left AV valves

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Anatomy of AVSD - biventricular AV connections

**Normal**

- RA
- LA
- RV
- LV

**AVSD**

- RA
- LA
- RV
- LV

**Four chamber section**

- **Common AV valve** (‘complete’ AVSD / AVSD with both ASD and VSD)

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Anatomy of AVSD - biventricular AV connections

Four chamber section

Separate AV valves

('partial' AVSD / primum ASD)

Subcostal

Parasternal

IBL= inferior bridging leaflet; SBL= superior bridging leaflet

LA
RA
LV
RV

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Anatomy of AVSD

AVSD

Common valve orifice (canal)

Separate valve orifices ('primum')

5-leaflet AV valve guards the common AV junction

Superior and Inferior Bridging Leaflets (joined or not)

SBL

IBL

NORMAL

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Anatomy of AVSD - the AV junction

NORMAL

Common valve orifice (AV canal)

Separate valve orifices (primum ASD)

AVSD

Normal
AVSD (5 leaflets)

Views from the apex

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Anatomy of AVSD - the left AV valve

Primum' or 'partial' AVSD

View from atria

View from ventricles

3 leaflets and 'cleft'

Papillary muscles

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Potential for shunting across AV Septal Defect
Number of valvar orifices within common AV junction
Relative sizes of chambers
Anatomy of AVSD - Levels of shunting
Anatomy of AVSD - Levels of shunting

Atrial septum

Coronary sinus

Inferior bridging leaflet

Superior bridging leaflet

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Anatomy of AVSD

Levels of shunting

Bridging leaflets attached to ventricular septum
Floating bridging leaflets
Bridging leaflets attached to atrial septum

interatrial
interatrial & interventricular
interventricular

Early presentation:

Magnitude of L to R shunting across interventricular communication (VSD component) depends both on size of the VSD and the ratio of pulmonary to systemic vascular resistance.

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Anatomy of AVSD - Levels of shunting

Bridging leaflets attached to atrial septum

interventricular

RA

RV

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Anatomy of AVSD – Inlet/Outlet lengths

Normal

Outlet

Inlet

scoop

Atrioventricular septal defect

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Anatomy of AVSD - scoop extension

Indexed values
• Depth: \( d/j \times 100 \) (%)
• Height: \( h/j \times 100 \) (%)
• LVOT diameter: \( l/J \times 100 \) (%)


16 / 31 hearts with complete AVSD

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16 / 31 'canal AVSD' specimens had antero-superior extension of scoop

- LVOT diameter did not correlate with scoop depth
- LVOT diameters were significantly narrower in hearts with A-S extension than without

A-S extension has more impact on LVOT than scoop depth

Anatomy of AVSD - AV conduction system

Branching bundle exposed on septal crest

Right ventricle

Right atrium

Coronary sinus

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Anatomy of AVSD - Associated malformations

- Right ventricular outflow obstruction
  - tetralogy of Fallot
- Left ventricular outflow obstruction
- Isomeric arrangement of the atrial appendages
  (heterotaxy); left-hand ventricular topology
  (‘ventricular inversion’)
- Chamber disproportion
- Deviation of ventricular septum
- others......persistent left sup caval vein, coarct., etc
Anatomy of AVSD - Associated malformations

Tetralogy of Fallot

Stenotic RV outlet

PT
SBL
RV

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Anatomy of AVSD - Associated malformations

LV outflow obstruction

LV

Papillary muscle

aneurysm

LV

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Relative size of chambers: dominance of one half

Dominant right, hypoplastic left heart

Anatomy of AVSD - Associated malformations

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Anatomy of AVSD - Associated malformations

Relative size of chambers: deviated ventricular septum

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Anatomy of AVSD

- A malformation characterised by a common AV junction and biventricular AV connections
- Associated malformations

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