Grown-up Congenital Heart Disease - How to follow the new ESC Guidelines:

Unoperated shunt lesions: when is closure indicated?

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Unoperated Shunt Lesions: When is closure indicated?

- Atrial septal defect (ASD)
- Ventricular septal defect (VSD)
- Atrioventricular septal defect (AVSD)
- Patent ductus arteriosus (PDA)
Atrial septal defect: When is closure indicated?

- RV volume overload

ASD                              NORMAL
Atrial septal defect: When is closure indicated?

- **Pulmonary overcirculation**

- **Increase in PAP with age, severe PVD rare (<5%)**

![Graph showing relationship between age and PAPsyst.]

<table>
<thead>
<tr>
<th>Age Group</th>
<th>PAPsyst. Mean ± S</th>
<th>PAPsyst. &gt;35 mmHg</th>
</tr>
</thead>
<tbody>
<tr>
<td>4th Decade</td>
<td>33 ± 9</td>
<td>43</td>
</tr>
<tr>
<td>5th Decade</td>
<td>35 ± 6</td>
<td>52</td>
</tr>
<tr>
<td>6th Decade</td>
<td>39 ± 11</td>
<td>50</td>
</tr>
<tr>
<td>7th Decade</td>
<td>40 ± 9</td>
<td>57</td>
</tr>
<tr>
<td>8th Decade</td>
<td>60 ± 17</td>
<td>100</td>
</tr>
</tbody>
</table>

\[ R = 0.65 \quad p < 0.0001 \]

Humenberger M......Baumgartner H
Eur Heart J 2010 in press
Atrial septal defect: Natural History

- Palpitations (atrial arrhythmias / afib)
- Reduced exercise capacity
- Shortness of breath
- Right heart failure
- Embolic events (afib, paradoxical emb.)
- Pulmonary infections
- Sudden death
- (chest pain)

Continuous increase of symptomatic pts. with age

Majority is symptomatic at age > 40 years
Atrial septal defect closure at age >40 yrs

Attie F et al   J Am Coll Cardiol 2001;38:2035-42
## Atrial septal defect closure at age >40 yrs

<table>
<thead>
<tr>
<th>Endpoint</th>
<th>Medical (n = 241)</th>
<th>Surgical (n = 232)</th>
<th>$p$ Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Totality of events</td>
<td>50 (20.7%)</td>
<td>26 (11.1%)</td>
<td>0.0046</td>
</tr>
<tr>
<td>Heart failure</td>
<td>9 (3.7%)</td>
<td>5 (2.1%)</td>
<td>0.3982</td>
</tr>
<tr>
<td>Pulm. embolism</td>
<td>4 (1.6%)</td>
<td>5 (2.1%)</td>
<td>0.8266</td>
</tr>
<tr>
<td>Periph. embolism</td>
<td>1 (0.4%)</td>
<td>3 (1.3%)</td>
<td>0.4056</td>
</tr>
<tr>
<td>Stroke</td>
<td>4 (1.6%)</td>
<td>6 (2.6%)</td>
<td>0.3727</td>
</tr>
<tr>
<td>Rec. pneumonia</td>
<td>24 (10.0%)</td>
<td>6 (2.6%)</td>
<td>0.0012</td>
</tr>
<tr>
<td>Sudden death</td>
<td>7 (2.9%)</td>
<td>2 (0.9%)</td>
<td>0.0837</td>
</tr>
<tr>
<td>Secondary</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Total mortality</td>
<td>14 (5.8%)</td>
<td>10 (4.3%)</td>
<td>0.1934</td>
</tr>
</tbody>
</table>

Median FU 7.3 (2-13) yrs

Attie F et al J Am Coll Cardiol 2001;38:2035-42
Atrial septal defect: Benefit of ASD closure in adults

Patients (%)

Pts < 40yrs  Pts 40 – 60yrs  Pts > 60yrs

Humenberger M......Baumgartner H
Eur Heart J 2010 in press
Atrial septal defect: Benefit of ASD closure in adults

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Systolic Pulmonary Artery Pressure (mmHg)

- < 40y
- 40-60y
- > 60y

sPAP pre, sPAP day 1, sPAP week 1, sPAP month 3
Atrial septal defect: Benefit of ASD closure in adults

Right Ventricular size (mm)

- RV size pre
- RV size day 1
- RV size week 1
- RV size month 3

- < 40y
- 40-60y
- > 60y

Humenberger M......Baumgartner H
Eur Heart J 2010 in press
Atrial septal defect: When is closure indicated?

- **ASD closure and severe PAH**
  - severe PVD is rare (<5%), young adults, ≈iPAH
  - benefit in pts. with PVR > 5 WU questionable
  - Pts. with PVR / PAP > 2/3 SVR / SP have in general no sign. L-R shunt and are unlikely to benefit from closure
  - Testing of vasoreactivity (preferably NO)
  - Targeted PAH therapy

- **ASD closure and severe LV dysfunction**
  - increase in preload may cause HF
  - Testing with balloon occlusion
Atrial septal defect closure

- Transcatheter ASD closure can be performed in appr. 85% of secundum ASDs with very low morbidity and almost no mortality.
- For surgical closure in the elderly, risk has to weighed against benefit.
Indications for Intervention in Atrial Septal Defect

Patients with significant shunt (signs of RV volume overload) and PVR < 5 WU should undergo ASD closure regardless of symptoms.

Device closure is the method of choice for secundum ASD closure when applicable.

All ASDs regardless of size in patients with suspicion of paradoxical embolism (exclusion of other causes) should be considered for intervention.

Patients with PVR ≥ 5 WU but < 2/3 SVR or PAP < 2/3 systemic pressure (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy) and evidence of net L-R shunt (Qp:Qs > 1.5) may be considered for intervention.

ASD closure must be avoided in patients with Eisenmerger physiology.

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a = class of recommendation. b = level of evidence.
ASD = atrial septal defect; L-R shunt = left-to-right shunt; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; SVR = systemic vascular resistance; WU = Wood units.
Ventricular Septal Defect: Presentation of adults

- **VSD considered to small for closure during childhood:**
  - **small:** normal LV (no volume overload), normal PAP, no symptoms
  - good outcome
  - **borderline or moderate:** LV volume overload (risk of late development of heart failure), elevated PAP
  - risk of late complications

- **Residual VSD after closure:** presentation dependent on defect size

- **VSD – Eisenmenger:** cyanosis (severe PVD, shunt reversal)

Gabriel H et al  J Am Coll Cardiol 2002
Soufflet V et al  Am J Cardiol 2010
Ventricular Septal Defect: When is closure indicated?

Careful evaluation with regard to:

- **LV volume overload** (echo, CMR)
- **Pulmonary vascular disease** (cardiac catherization with assessment of PAP and PVR including vasoreactivity testing – preferably with NO – in case of high values)
- **Response to exercise:**
  - exercise capacity etc.
  - desaturation with exercise in patients with PAH
- **Aortic regurgitation**
Doubly committed VSD with AV prolapse causing aortic regurgitation
Indications for Intervention in Ventricular Septal Defect

Patients with symptoms that can be attributed to L-R shunting through the (residual) VSD and who have no severe pulmonary vascular disease (see below) should undergo surgical VSD closure.

Asymptomatic patients with evidence of LV volume overload attributable to the VSD should undergo surgical VSD closure.

Patients with a history of IE should be considered for surgical VSD closure.

Patients with VSD associated prolapse of an aortic valve cusp causing progressive AR should be considered for surgery.

Patients with VSD and PAH should be considered for surgery when there is still net L-R shunt (Qp:Qs > 1.5) present and PAP or PVR are < 2/3 of systemic values (baseline or when challenged with vasodilators, preferably nitric oxide, or after targeted PAH therapy).

Surgery must be avoided in Eisenmerger VSD and when exercise induced desaturation is present.

If the VSD is small, not subarterial, does not lead to LV volume overload or pulmonary hypertension and there is no history of IE, surgery should be avoided.

a = class of recommendation. b = level of evidence.

AR = aortic regurgitation; IE = infective endocarditis; LV = left ventricle; PAH = pulmonary arterial hypertension; L-R shunt = left-to-right shunt; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; VSD = ventricular septal defect.

www.escardio.org/guidelines
Atrioventricular Septal Defect

Partial

Intermediate

Complete

www.escardio.org/guidelines
# Indications for Intervention in Atrioventricular Septal Defect

<table>
<thead>
<tr>
<th>Complete AVSD</th>
<th>Partial AVSD</th>
<th>AV valve regurgitation</th>
<th>SubAS</th>
</tr>
</thead>
<tbody>
<tr>
<td>- Cardiac surgery must be avoided in patients with Eisenmenger physiology. In case of doubt, PVR testing is recommended. For indication of intervention see also VSD.</td>
<td>- Surgical closure should be performed in case of significant volume overload of the RV. For further details see ASD.</td>
<td>- Symptomatic patients with moderate to severe AV valve regurgitation should undergo valve surgery, preferably AV valve repair.</td>
<td>- See LVOTO</td>
</tr>
<tr>
<td>- Asymptomatic patients with moderate or severe left-sided valve regurgitation and LVESD &gt; 45 mm and/or impaired LV function (LVEF &lt; 60%) should undergo valve surgery when other causes of LV dysfunction are excluded.</td>
<td>- Surgical repair should be considered in asymptomatic patients with moderate or severe left-sided AV valve regurgitation who have signs of volume overload of the LV and a substrate of regurgitation that is very likely to be amenable for surgical repair.</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

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**Class**

<table>
<thead>
<tr>
<th>Class</th>
<th>Level</th>
</tr>
</thead>
<tbody>
<tr>
<td>III</td>
<td>C</td>
</tr>
<tr>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>I</td>
<td>C</td>
</tr>
<tr>
<td>IIa</td>
<td>C</td>
</tr>
</tbody>
</table>

**a = class of recommendation. b = level of evidence.**

ASD = atrial septal defect; AV = atrioventricular; AVSD = atrioventricular septal defect; LV = left ventricle; LVEF = left ventricular ejection fraction; LVESD = left ventricular end-systolic diameter; PVR = pulmonary vascular resistance; RV = right ventricle; SubAS = subaortic stenosis; VSD = ventricular septal defect.
**Patent Ductus Arteriosus:**

**Presentation of adults**

- **Small PDA:** normal LV (no volume overload), normal PAP, no symptoms

- **Moderate PDA with predominant LV volume overload:** Enlarged LV with normal or reduced LV function (left heart failure)

- **Moderate PDA with predominant PAH:** Pressure loaded RV (right heart failure)

- **PDA – Eisenmenger:** differential cyanosis
Small PDA
Moderate PDA – LV volume overload
Moderate PDA – RV Pressure overload
Indications for Intervention in Patent Ductus Arteriosus

PDA should be closed in patients with signs of LV volume overload

PDA should be closed in patients with PAH but PAP < 2/3 of systemic pressure or PVR < 2/3 of SVR

Device closure is the method of choice where technically suitable

PDA closure should be considered in patients with PAH and PAP > 2/3 of systemic pressure or PVR > 2/3 of SVR but still net L-R shunt (Qp:Qs > 1.5) or when testing (preferably with nitric oxide) or treatment demonstrates pulmonary vascular reactivity

Device closure should be considered in small PDAs with continuous murmur (normal LV and PAP)

PDA closure should be avoided in silent duct (very small, no murmur).

PDA closure must be avoided in PDA Eisenmerger or patients with severe PAH and exercise-induced lower limb desaturation

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a = class of recommendation. b = level of evidence.
L-R shunt = left-to-right shunt; LV = left ventricle; PAH = pulmonary arterial hypertension; PAP = pulmonary artery pressure; PDA = patent ductus arteriosus; PVR = pulmonary vascular resistance; Qp:Qs = pulmonary to systemic flow ratio; SVR = systemic vascular resistance.

www.escardio.org/guidelines
Unoperated Shunt Lesions: When is closure indicated?

- Shunt closure is indicated in the majority of adults with atrial septal defects (frequent) and in selected patients ventricular septal defect, AVSD and patent ductus arteriosus.
- Management decision requires careful assessment of ventricular volume overload and of the pulmonary circulation in experienced hands.
- Recommendations are mainly based on expert consensus (evidence level C).
Thank you for your attention!
Atrial Septal Defect: Diagnostic Work-up

- **Echocardiography:** key diagnostic technique providing diagnosis and quantification (RV volume overload!)
  - TEE for precise evaluation of secundum defects prior to device closure (size, residual septum´s morphology, rim size and quality, exclusion of additional defects and confirmation of normal pulmonary venous connection) and of sinus venosus defects
  - Other key info to be provided includes PAP and TR

- **CMR/CT:** if echo is insufficient, particularly for RV volume overload and pulmonary venous connection

- **Cardiac catheterisation:** estimation of PVR when echo PAP >50% of systemic pressure
Atrial Septal Defect: Follow-up

- **FU evaluation** should include assessment of a residual shunt, RV size and function, TR and PAP by echocardiography as well as of arrhythmias by history, ECG and only if indicated (not routinely) Holter.

- Late post-op. **arrhythmias** after surgical repair at age <40 years are most frequently intraatrial reentrant tachycardia or atrial flutter which can be successfully treated with radiofrequency ablation. Without repair or with repair after 40 years, atrial fibrillation becomes more common (oral anticoagulation!)

- Pts. repaired <25yrs without relevant sequelae or residuae do not require regular FU; the others should be followed on a regular basis including evaluation in specialised GUCH centers.
Ventricular Septal Defect: Diagnostic Work-up

- **Echocardiography:** key diagnostic technique providing diagnosis and quantification (LV volume overload!) Key findings to provide are location, number and size of defects, severity of LV volume overload and PAP. Check for AR due to prolapse of the right or non-coronary cusp (especially in outlet/supracristal and high perimembranous VSDs) and for DCRV.

- **CMR:** if echo is insufficient, particularly for assessment of LV volume overload and shunt quantification.

- **Cardiac catheterisation:** estimation of PVR when echo PAP >50% of systemic pressure
Ventricular Septal Defect: Follow-up

● **FU evaluation** should include assessment of AR, TR, degree of (residual) shunt, LV dysfunction, elevation of PAP, development of DCRV and development of discrete subaortic stenosis by echocardiography.

● Possible development of complete AV block requires attention (patients who develop bifascicular block or transient trifascicular block after VSD closure are at risk).

● Patients with LV dysfunction, residual shunt, PAH, AR, RVOT or LVOT obstruction should be seen every year, small VSDs in 3-5-year intervals; After device closure: regular follow-up until 2 years and then depending on the result every 2-4 years is recommended. After surgical closure without residual abnormality 5-year intervals.
Atriventricular Septal Defect (AVSD): Diagnostic Work-up

- **Echocardiography:** key diagnostic technique providing assessment of each anatomic component of the AVSD, of the AV-valves and their connections (straddling; overriding) and the severity and exact substrate of AV valve regurgitation, the magnitude and direction of intracardiac shunting, LV and RV function, PAP and the assessment of presence/absence of sub-aortic stenosis.

- **CMR:** indicated when additional quantification of ventricular volumes and function or intracardiac shunting is required for decision making.

- **Cardiac catherisation:** estimation of PVR when echo PAP >50% of systemic pressure
AVSD: Follow-up

- **FU evaluation** should pay particular attention to residual shunt, AV valve malfunction, LV and RV enlargement and dysfunction, PAP elevation, subaortic stenosis and arrhythmias.

- Life-long regular follow-up of all patients, operated and unoperated, with an AVSD is recommended including evaluation in specialised GUCH centers. The frequency of outpatient visits depends on the presence and severity of residual abnormalities. A surgically repaired AVSD without significant residual abnormalities should be seen at least every 2 to 3 years. In case of residual abnormalities, the intervals should be shorter.
Patent Ductus Arteriosus (PDA): Diagnostic Work-up

- **Echocardiography:** key diagnostic technique providing the diagnosis (may be difficult in pts. with Eisenmenger physiology), the degree of LV volume overload, PAP, PA size and right heart changes.

- **CMR:** indicated when additional quantification of LV volumes or evaluation of PA anatomy are required.

- **Cardiac catherisation:** estimation of PVR when echo PAP >50% of systemic pressure
Patent Ductus Arteriosus: Follow-up

- **FU evaluation:** echocardiographic evaluation should include LV size and function, PAP, residual shunt and associated lesions.
- Patients with no residual shunt, normal LV and normal PAP do not require regular follow-up after 6 months.
- Patients with LV dysfunction and patients with residual PAH should be followed with 1-3 years intervals depending on severity including evaluation in specialised GUCH centers.