The ABCs of ASDs

John M. Lasala MD PhD
Director Interventional Cardiology and Structural Heart Disease
Professor of Medicine
Washington University
St Louis Mo
ATRIAL SEPTAL DEFECT

GENERAL PHYSIOLOGY

- RA and RV volume overload
- Excess volume may result in pulmonary hypertension may develop
  - More often in women
  - Generally not severe
- May result in paradoxical emboli

Brickner et al. *NEJM* 2000
Hemodynamic Consequences of ASD

• Magnitude of and direction of flow depends on
  – Size of the defect
  – Relative diastolic filling properties of the left and right ventricles.
    • Increased left-to-right shunting results from reduced LV compliance (eg, LVH) and mitral stenosis.
    • Reduced left-to-right shunt and/or reversal of shunt (right-to-left shunt) results from reduced RV compliance (eg, pulmonary hypertension or pulmonary stenosis) and tricuspid stenosis

Webb G and Gatzoulis MA. Circulation 2006;114;1645-1653
Hemodynamic Consequences of ASD

• Size and Shunt
  – As a rule, an ASD must be at least 10 mm in diameter to carry a significant left-to-right shunt
    • CAVEAT: symptoms may develop with increasing age even with small defects owing to an increase in shunting caused by a decrease in LV compliance secondary to coronary artery disease, acquired valvular disease, or hypertension.
  – A left-to-right atrial shunt is considered significant when the Qp/Qs ratio is greater than 1.5/1.0, or if it causes dilation of the right heart chambers.

• Chronic volume overload of the pulmonary vasculature may result in pulmonary arterial hypertension

Webb G and Gatzoulis MA. Circulation 2006;114;1645-1653
Sinus Venosus
ASD: IVC Type
Sinus Venosus ASD: IVC Type

Note: ASD with PFO
Secundum ASD

- Atrial septal defects
  - Three-dimensional size
  - Assessment of rim
    - Percutaneous closure
  - Assessment of anomalous pulmonary veins
Rationale for Intervening

- Natural history of ASD diagnosed in childhood is that the ASD diameter when untreated increases in 65% of cases, and 30% will have more than a 50% increase in diameter. Only 4% of ASDs close spontaneously.

- Although small ASDs of <5 mm and no evidence of RV volume overload do not impact the natural history of the individual and thus may not require closure:
  - Paradoxical embolism may occur
  - Some small defects however may have progressive increase in left-to-right shunt depending on LV and LA pressures

  - 200 unoperated patients >40 yo followed 1.6 to 22 years.
  - 37 events (18.5%): 5 sudden death, 7 heart failure, 13 severe pulmonary infection, 5 embolisms and 4 strokes.
  - Predictors of poor outcome were age at presentation, pulmonary HTN or arterial O2 sat < 80%

Rationale for Intervening

- Larger defects with evidence of RV volume overload on echocardiography usually only cause symptoms in the third decade of life, and closure is usually indicated to prevent long-term complications.

### Definite and Potential Benefits of ASD Closure

<table>
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<tr>
<th>Benefit</th>
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<tr>
<td>RV and RA size ↓</td>
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<tr>
<td>LV size ↑</td>
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<tr>
<td>PA pressure ↓</td>
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<tr>
<td>Right-to-left shunting and embolism ↓</td>
</tr>
<tr>
<td>Exercise capacity ↑</td>
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<tr>
<td>NYHA class ↓</td>
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<tr>
<td>Atrial arrhythmias ↓</td>
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Nomenclature of ASD Rims

- Conventionally, the 5 rims of a secundum ASD are labeled as:
  - Aortic (anterosuperior)
  - Atrioventricular (AV) valve (mitral or anteroinferior)
  - Superior venacaval (SVC or posterosuperior)
  - Inferior venacaval (IVC or posteroinferior)
  - Posterior (from the posterior free wall of the atria).

By conventional definition:
1. Margin $\geq 5$ mm is considered adequate
2. Margin $\leq 3$ mm is considered absent
Secundum ASD Rims

Secundum ASD rims: 1. SVC  2. Aortic  3. AV  4. IVC  5. RPV

Deficient aortic rim

Deficient IVC rim

Deficient SVC rim

Deficient RPV rim

Currently available devices within the United States for percutaneous closure of atrial-level defects within randomized controlled trials.

- **CardioSEAL™**
  (NMT Medical; Massachusetts, USA)

- **STARFlex™**
  (NMT Medical; Massachusetts, USA)

- **Amplatzer PFO Occluder™**
  (AGA Medical, Minnesota, USA)

- **Helex™ Septal Occluder**
  (W.L. Gore and Associates; Delaware, USA)

- **Cardia PFO Occluder (Intracept™)**
  (Cardia; Minnesota, USA)

- **Premere PFO Closure™**
  (St. Jude Medical, Minnesota, USA)

Currently available FDA approved devices in the United States

**AMPLATZER ASO**
Polyester material sewn into 0.004”-0.008” braided nitinol wires

**GORE® HELEX® Septal Occluder**
ePTFE patch material supported by a single nitinol wire frame

*Inglessis I, Landzberg M. Circulation 2007;115:1622-1633*
Fenestrated Secundum ASD
Failure to Close a Fenestrated ASD
Atrial Septal Defect

- ASD can be associated with additional malformations in nearly 30% of cases
  - AV septal defects (primum ASD) → cleft in the anterior mitral valve leaflet
  - Sinus venosus defects  frequently (rarely with secundum ASD) → partial anomalous venous drainage of the right pulmonary veins
  - Any ASD → Mitral valve prolapse
  - Any ASD → Valvular pulmonic stenosis
  - Coronary sinus septal defect → partial or total anomalous pulmonary venous connection and/or a persistent left superior vena cava draining to the coronary sinus.

SECUNDUM ASD - ASSOCIATED DEFECTS

NORMAL

LEFT INNOMINATE VEIN INJECTION (Straight AP)

NORMAL?
SECUNDUM ASD - ASSOCIATED DEFECTS
SECUNDUM ASD - ASSOCIATED DEFECTS:

Partial Anomalous Pulmonary Venous Return to the Left Innominate Vein

LPA Angio (LAO) showing “split” drainage of left pulmonary veins
OTHER DEFECTS ASSOCIATED WITH SECUNDUM ASD TO WATCH FOR:

- PAPVR
  - Left Upper Pulmonary Vein to LSVC draining to innominate ("bridging") vein
  - Right Pulmonary Veins to the SVC and/or the backwall of the RA

- Pulmonary Valve Stenosis
  Balloon Valvuloplasty PSG >40mmHg
SECUNDUM ASD - ASSOCIATED DEFECTS
SECUNDUM ASD- ASSOCIATED DEFECTS

RIGHT PARTIAL ANOMALOUS PULMONARY VENOUS RETURN:

- RUPV to SVC-RA JUNCTION
- RLPV to BACKWALL of RIGHT ATRIUM
SECUNDUM ASD VARIANTS:

“FOSSA ASD”

Also known as “valve incompetent” PFO
SECUNDUM ASD VARIANTS:
MULTI-FENESTRATED TYPES

...they are **not** all the same!
SECUNDUM ASD VARIANTS:

ANEURYSMAL ("windsock")
FENESTRATED TYPE
Definition of Complex Secundum ASDs

- Present in 28% of cases (Pedra, et al. JIC 2004;16;117)
- >26mm stretch diameter with a deficient rim < 4mm
- 2 separate ASDs with a distance > 7mm
- Fenestrated atrial septum
- Redundant or hypermobile septum
So when is a paucity/absence of rim really an issue?

<table>
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<tr>
<th>Type of Rim Absence</th>
<th>Acceptable Device Candidate</th>
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<tbody>
<tr>
<td>Retro-Aortic Rim Absence</td>
<td>Yes</td>
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<td>Posterior Rim Absence</td>
<td>Probably NO; depends on defect size (enhanced migration risk)</td>
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<td>Inferior Rim</td>
<td>Depends on extent &amp; C.S. *</td>
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<td>Superior Rim</td>
<td>Yes, except true S.V. + PAPVR</td>
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<tr>
<td>AV Valve Rim</td>
<td>Depends on overall extent &amp; proximity of device to M.V. leaflet *</td>
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POOR ANATOMIC ASD CANDIDATES:

- Absence of significant posterior rim in defect larger than 1.5 cm:
  - Increased risk of device migration
Large ASDs: closure methods

- Rotation on delivery sheath
- Deployment in RUPV
- Deployment in LUPV
- Use of alternative sheaths (Mullins, Hausdorf, modified Amplatzer)
- Use of right coronary catheter
- Balloon assisted technique
- Use of dilator as buttress
Courtesy of Evan M. Zahn, MD, Miami Children’s Hospital
Kannan, Catheter Cardiovasc Intervent, 2003
ASD DEVICE TIPS & TRICKS

Hausdorf Sheath (COOK Corp.)

Tip: 3-D curve that keeps sheath more perpendicular to ASD plane when there is an anterior-superior ASD with absent retro-aortic rim

Available in 10-12 French Sizes
ADVANCED ASD CLOSURE

Sheath Tips and Tricks:

NEW AGA TorqVue® Sheath

Hausdorf Sheath (Cook, Inc.)
ASD: no rim
Challenging Case: Large ASD
Large ASD

18 y/o female referred for evaluation of asymptomatic heart murmur

Transthoracic echo confirms presence of large ASD with dilation of RA and RV

CXR with mild cardiomegaly and shunt vascularity
Large ASD

- Cardiac cath demonstrates normal right heart pressures and Qp:Qs 3:1
- ICE demonstrates large secundum ASD
Large ASD
Large ASD

- Several unsuccessful attempts were made to deploy 28 mm Amplatzer device in conventional manner
- Attempts at left PV deployment technique also unsuccessful secondary to prolapse of LA disc through retro-aortic rim
Large ASD
Large ASD
Large ASD
Large ASD
Large ASD
Large ASD
Large ASDs with Deficient Anterosuperior Rims

- Largest percentage of deficient rims (42%)
- Amplatzer discs must straddle the aorta (riding bareback)
- Keep discs flared around the aorta to prevent erosion
- Avoid using a device > 1.5 times the TEE/ICE diameter
- Watch for encroachment on structures- MV, SVC, RUPV
- Know the tricks presented for appropriate placement
Closure of Multiple ASDs
Multiple ASDs

- Occurs in up to 6.6% of ASDs
- May require multiple devices to close the defects if far enough apart
- Single device may be adequate for closely spaced defects
- Occasionally balloon sizing will convert multiple defects into a single defect
Multiple ASDs

- Most fenestrated defects can be closed with the cribiform device
- Multiple ASDs may require more than 1 device
  - If defects $\geq 7$ mm apart 2 devices (ASO) may be implanted
    - May implant sequentially or simultaneously
Multiple ASDs
Multiple ASDs
Multiple ASDs
Multiple ASDs
2 ASDs
Fenestrated ASD (multiple holes)

Courtesy Y. Joe Woo, MD
HUP Cardiac Surgery
Case Presentation- ASD closure Due to Hypoxia

• 34 yo woman with a long h/o SOB
  • Had difficulty playing sports as a child
  • Severe SOB with each of her 2 pregnancies
  • Once given dx of asthma (not treated)

• More recently, c/o of SOB with 1 flight of stairs, carrying her baby, and also associated with some dizziness and lightheadedness

• Work-up including a TTE which suggested a left to right shunt.
- Secundum ASD measured 0.5 cm diameter
- Qp:Qs ratio of 1.4
- Referred for percutaneous ASD closure
Case Presentation

- Admitted for RHC and ICE with planned ASD closure
  - Qp:Qs only 1.1:1
  - Normal Pa pressure
ICE contrast study

Rest

After arm exercise

Washington University in St. Louis
School of Medicine
Hypoxia in Patients with Interatrial Septal Defects (IASD)

- Can be persistent, intermittent, or positional
- Mechanism involves transient or persistent elevation in RAP>LAP, or redirection of IVC blood flow toward septum
- Diagnosis can be challenging
  - Requires documentation of R-to-L shunt while hypoxemic
  - Confirmed by improvement in hypoxia after closure
- Associated with a wide variety of conditions
  - Pulmonary AVM
  - Liver Disease
  - Chronic Lung Disease
  - Amiodarone Toxicity
  - Pulmonary Emboli
  - Aortic Aneurysm
  - Hypovolemia
  - Positive Pressure Ventilation
  - Post-pneumonectomy
  - RV Infarction
  - Cardiopulmonary Bypass
Saturation (%)

PRE  POST

87%  96%

P=0.002

Ilkhanoff and Herrmann  J Int Card 2005;18:227-32
Therapeutic Intervention

- Closure performed
  - #25 AGA Cribiform ASO inserted in PFO
  - #14 AGA Atrial septal occluder for ASD
• In follow-up, all of her symptoms have resolved
ASD History:

- Recent murmur appreciated and echo revealed a large secundum ASD with estimated diameter of 27 and 28mm. R-sided chambers were enlarged, mod Pulm HTN was present. MRI ruled out sinus venosus defect with any anomalous pulmonary drainage.

- He underwent initial attempt at percutaneous closure at OSH, but defect unsuccessfully closed. Patient referred to Barnes-Jewish for second attempt at percutaneous closure.
Cath Films - ASD Closure:
Case Report cont:

ASD Procedure:

- Fluoroscopic and TEE guidance used.
- 38mm Amplazter device used for procedure with successful deployment.
- Bubble study negative immediately post-procedure.
Cath Films – ASD Closure:
Cath Films – ASD Closure:
TEE Post Deployment
Later the Same Day…

- Patient was transferred to the floor
- No complaints
- Routine TTE obtained that night per protocol
- 911 page sent
Echo Post Procedure
Echo post-procedure:
Deficient Posteroinferior Rims

- A real challenge as these tend to be larger ASDs
- MD beware if rim is < 3mm.
- Increased complication rate- IVC or Pulmonary vein obstruction, encroachment on mitral valve, frank embolization
The Future of Imaging for Structural Heart Disease
3D Imaging: Acunav

10 Fr system
### ACC/AHA Guidelines

#### Class 2.5.2. Recommendations for Interventional and Surgical Therapy

<table>
<thead>
<tr>
<th>Level of Evidence</th>
<th>I 1.</th>
<th>IIa 1.</th>
<th>IIb 1.</th>
<th>III</th>
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<tr>
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<td>Closure of an ASD either percutaneously or surgically is indicated for right atrial and RV enlargement with or without symptoms.</td>
<td>Surgical closure of secundum ASD is reasonable when concomitant surgical repair/replacement of a tricuspid valve is considered or when the anatomy of the defect precludes the use of a percutaneous device.</td>
<td>Closure of an ASD, either percutaneously or surgically, may be considered in the presence of net left-to-right shunting, pulmonary artery pressure less than two thirds systemic levels, PVR less than two thirds systemic vascular resistance, or when responsive to either pulmonary vasodilator therapy or test occlusion of the defect (patients should be treated in conjunction with providers who have expertise in the management of pulmonary hypertensive syndromes).</td>
<td>Patients with severe irreversible PAH and no evidence of a left-to-right shunt should not undergo ASD closure.</td>
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<td>A sinus venosus, coronary sinus, or primum ASD should be repaired surgically rather than by percutaneous closure.</td>
<td>Closure of an ASD, either percutaneously or surgically, is reasonable in the presence of: a. Paradoxical embolism. b. Documented orthodeoxia-platypnea.</td>
<td>2. Concomitant Maze procedure may be considered for intermittent or chronic atrial tachyarrhythmias in adults with ASDs.</td>
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<td>Surgeons with training and expertise in CHD should perform operations for various ASD closures.</td>
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