Case Presentation
R.G. was a 5 year old female patient with Ebstein’s malformation of the tricuspid valve. She initially presented at age of 10 months for evaluation of an asymptomatic heart murmur. Her history was unremarkable and she had no symptoms or signs of congestive heart failure or arrhythmia. She had shown normal growth and development. Physical examination revealed a well-appearing child with normal vital signs, normal pulses in all extremities, and a prominent right ventricular impulse on palpation of the precordium. On auscultation, the first heart sound was normal and the second sound showed a wide fixed split. There was no S3 or S4, but some indistinct systolic clicks were heard over the lower precordium. A grade 2/6 soft ejection systolic murmur was heard best at the left upper sternal border without radiation. An electrocardiogram showed normal sinus rhythm with normal intervals, a rightward QRS axis of 155º, with an RSR’ pattern in V1 and V2 and a deep S in V6, suggestive of right ventricular enlargement. An echocardiogram showed normal ventricular function, with moderate dilation of the right ventricle. There was mild to moderate Ebstein’s malformation of the tricuspid valve, characterized by dysplasia and inferior displacement of the leaflets of the valve, but no evidence of obstruction and only trivial regurgitation. However, a secundum atrial septal defect (ASD) measuring 1.1 cm in diameter, with a good circumferential rim of atrial septum was noted. Flow across the defect was bidirectional. There was no evidence of pulmonary hypertension, and no other abnormalities were detected. She underwent a cardiac catheterization. At the start of the procedure a transesophageal echocardiogram was performed which confirmed the anatomy, and hemodynamic assessment confirmed suitability for closure of the ASD (Figure 1A). A 20 mm double umbrella device (CardioSEAL) was placed for closure of the ASD, with a small residual shunt noted with the transesophageal echocardiogram (Figure 1B). A three-dimensional echocardiogram showed the ASD device pre and post release (Figure 2). The patient made a good recovery and had ongoing follow-up. However, at 8 years of age she collapsed at school. Paramedics documented supraventricular tachycardia at a rate of 260 bpm, and on cardioversion she developed ventricular fibrillation from which she could not be resuscitated. An autopsy was performed.

Stuff It or Stitch It:
Innovations in the Closure of Atrial Septal Defects

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Brian McCrindle, M.D.
Atrial Septal Defects

Embryologically, the major septa of the human heart are formed between the 27th and 37th days of development. The formation of the atrial septum involves two actively growing masses of tissue (septum primum and septum secundum). Apoptosis produces perforations in the upper portion of the septum primum, creating the ostium secundum. The septum secundum never forms a complete partition in the atrial cavity, leaving an opening called the foramen ovale. This elongated cleft between the two atrial cavities allows passage of blood from the right atrium to the left side in utero. After birth, when pulmonary circulation and pulmonary venous return increases, pressure in the left atrium increases and the valve of the foramen ovale is pressed against the septum secundum, functionally closing the oval foramen and separating the right and left atria.

An atrial septal defect (ASD) may be located at various sites in the septum, most commonly in the septum secundum. It is twice as common in females than in males and has an incidence of 6.4 per 10,000 births. Four types of ASDs exist: secundum defect, primum defect, sinus venosus defect and coronary sinus defect (Figure 3). Ostium secundum defect is the most common type of ASD, accounting for 75% of all ASDs. This ASD is located in the central portion of the septum (fossa ovalis) and is formed by excessive resorption of the septum primum or deficiency of the septum secundum. This defect may present as a normal variant with persistent patency of the foramen ovale (PFO), which is also located in the middle portion of the atrial septum, usually closes spontaneously with time, but remains probe-patent in 20 to 25% of individuals throughout life. Recent evidence has associated the presence of PFO in patients with stroke with an increased risk of recurrence. Primum ASDs account for 15% of all ASDs and are seen in the lower portion of the septum just above the atrioven-
of right heart structures and the lungs. On auscultation, a murmur may be heard; however it is not caused by the shunt at the atrial level. The heart murmur noted with ASD originates from the pulmonary valve and right ventricular outflow tract, and results from the increased blood flow passing through this normal-sized valve. Therefore, the murmur is systolic in timing (usually grade 2-3/6 systolic ejection murmur) and is maximally heard at the upper left sternal border. When the left-to-right shunt is large enough, a diastolic flow rumble may be audible at the lower left sternal border, representing increased flow across a normal tricuspid valve. With the increase in pulmonary blood flow from the shunt, pulmonary valve closure is delayed and the second heart sound is typically widely split and fixed.

Cardiomegaly with right ventricular (RV) and right atrial enlargement as well as a dilated pulmonary artery may be seen on chest x-ray. Echocardiography, which is diagnostic, may show increased RV dimension and paradoxical motion of the interventricular septum, which results from the RV volume overload. In older children and adolescents, optimal imaging of the atrial septum may not be possible with the ordinary transthoracic echocardiographic study, and transesophageal echocardiography (TEE) may be used as an alternative.

An electrocardiogram (ECG) of a child with an ASD may show signs of RV enlargement. The dilated RV cavity prolongs the time required for depolarization of the RV because of its longer pathway, producing an appearance of partial or complete right bundle branch block (RBBB) (with RSR’ in V1) in the ECG. The RBBB pattern in children with ASDs is not the result of actual block in the right branch of the His bundle. If the duration of the QRS complex is not abnormally prolonged, the ECG may be read as mild right ventricular hypertrophy (RVH). Right axis deviation of +90 to +180 degrees may also be seen on ECG.

Natural History

Secundum ASDs usually go undetected in the first year or two of life because of the lack of symptoms. A soft systolic murmur is the common reason for referral. Symptoms become more apparent in adolescents. Pulmonary vascular obstructive disease with serious pulmonary hypertension is rare, but may begin to make its appearance in the early twenties. Atrial arrhythmias (flutter or fibrillation) may occur in adults and is closely linked to the onset of congestive heart failure (CHF). CHF rarely develops in infancy.

Spontaneous closure of important secundum ASDs is rare beyond the first two years of life, although it has been reported. Brassard et al reported that 57% of the study population had spontaneous closure after 18 months of age, and 40% after age of 5 years. The greatest predictor of spontaneous closure is initial ASD size at diagnosis. Closure seems to occur significantly earlier in infants whose openings are <3mm in diameter. In a study by Senocak et al, 100% of the ASDs <3mm spontaneously closed at the end of the 18th month of life, while it took 4 months for 86% and 63% of the 3-5mm and 5-7 mm ASDs, respectively, to close. It is hypothesized that the presence of an atrial septal tissue flap originating from anatomical septum primum may relate to the mechanism of spontaneous closure. One study showed that 65% of defects actually increase in diameter over a mean study period of 3 years, at a mean rate of 0.8mm/year. In approximately 30% of patients, the ASD increased in size by 50% or more.

Management

Few patients with ASDs will have signs or symptoms of congestive heart failure, and therefore do not require medical management. Exercise restriction is unnecessary and prophylaxis for infective endocarditis is not usually indicated. In the rare infant with congestive heart failure, medical management is recommended, and can ameliorate symptoms and signs while the defect undergoes spontaneous closure.

It has been reported that surgical closure of the ASD had no impact on outcomes in adults when compared to medical therapy only. Conversely, Murphy et al concluded that surgery before age 25 was associated with improved outcomes. Further studies have confirmed this observation. The controversy remains in the optimal age and method of closure. There does not appear to be any significant benefit in ASD closure before 3 to 5 years of age in small to moderate sized asymptomatic defects. Long-term outcomes in patients who had closure before 15 years of age have been reported as good, however there was a high incidence of arrhythmias, with some patients having persistence of right ventricular dilation. Other studies have indicated that while there may be ongoing spontaneous closure in childhood, during a mean follow-up of 11.5 years, the size of the defect may increase. In a later editorial, it was concluded that large defects should be closed in childhood, while smaller defects required follow-up, and asymptomatic defects diagnosed in later adulthood may not require closure.

Surgical Closure

Surgical defect closure is traditionally performed while on cardiopulmonary bypass, through a median sternotomy incision with closure by either a simple suture or a pericardial or Teflon patch. Outcomes in children having had surgical closure have been studied and it was noted that immediate residual shunts were evident in 15%, decreasing to 8% for patients having >4 months follow-up. There were no deaths, but frequent complications occurred (Table 1).

<table>
<thead>
<tr>
<th>Table 1</th>
<th>Reported Complications Associated with Surgery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pneumothorax</td>
<td>Sepsis</td>
</tr>
<tr>
<td>Post-operative bleeding</td>
<td>Post-pericardiotomy syndrome</td>
</tr>
<tr>
<td>Pleural effusions</td>
<td>Chylothorax</td>
</tr>
<tr>
<td>Pericardial effusions</td>
<td>Post-operative pain</td>
</tr>
<tr>
<td>Thrombosis</td>
<td>Scarring, adhesions</td>
</tr>
<tr>
<td>Stroke</td>
<td>Phrenic nerve injury</td>
</tr>
<tr>
<td>Wound infection, mediastinitis</td>
<td></td>
</tr>
</tbody>
</table>

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Minimally invasive cardiac surgical techniques, with smaller skin incisions, have become popular for cosmetic reasons as well as improving comfort and recovery outcomes of surgery. A midline short transxiphoid incision with minimal sternal split has been reported in 10 patients (mean age: 5 years), with good feasibility and outcomes.17 The mini-sternotomy technique has been widely used; however no randomized studies have been performed to evaluate patient outcomes. In a non-randomized study, Laussen et al18 noted that mini-sternotomy patients had lower post-operative pain scores (not statistically significant) and no difference in length of ICU and hospital stay. At present, the only benefit of this technique appears to be an improved cosmetic result, while further studies need to assess its effect on pain reduction, hospital stay, or surgical stress.

Transcatheter Device Closure
The first device used to close an ASD was a plastic button developed in 1959,19 which could be implanted in the atrial septum via a thoracotomy. The first transcatheter closure of an ASD was reported by King et al20. This device was composed of two umbrella shaped discs of Teflon coated stainless steel and polyester fabric. In 1983, Rashkind21,22 developed a single disc device to close ASDs. This disc consisted of polyurethane foam on a wire umbrella-like skeleton, closing the defect by embedding in the atrial septum three small sharp “fish hooks” at ends of the wire skeleton. At that time, results of device implantation were mixed.22,23 This was followed by development of a double disk device which sandwiched the atrial septum. All further devices have adopted this basic design. These early successes have led to ongoing efforts to develop the ideal device (Table 2).

Table 2

<table>
<thead>
<tr>
<th>Feature</th>
<th>During Closure</th>
<th>After Successful Closure</th>
<th>After Failed Closure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small introducing catheter</td>
<td></td>
<td>Device lies flat against septum with a low profile</td>
<td>Embolization produces minimal hemodynamic stress</td>
</tr>
<tr>
<td>Adjustable and retrievable prior to release</td>
<td></td>
<td>Material that promotes full endothelialization</td>
<td>Easily retrievable after release</td>
</tr>
<tr>
<td>Flexible self-centering mechanism</td>
<td></td>
<td>Reliable device integrity until full endothelialization</td>
<td></td>
</tr>
<tr>
<td>Adjusts to variable septal topography</td>
<td></td>
<td>Minimal overlap to protect adjacent cardiac structures</td>
<td></td>
</tr>
</tbody>
</table>

Figure 4 summarizes the currently available devices being used either in clinical trials or in general use. The most promising device being used is the Amplatzer device (Figure 5), which is relatively easy to place, with a reported low incidence of residual leaks and possibly a greater degree of feasibility for device closure.24,25 A major advantage is that after the device has been partially or even fully opened, it may be readily withdrawn back into the delivery sheath, thus allowing multiple attempts at closure with the same device.26

Assessment of suitability for transcatheter closure is best done by transthoracic (TTE) and transesophageal (TEE) echocardiography.27 Current common echocardiographic criteria for feasibility for device closure include maximal ASD diameter of 20 mm, a septal rim >4 mm around at least 75% of the defect, safe distance (4-5 mm) from other structures (atrioventricular valves, vena cava, right pulmonary veins and coronary sinus), and patient size >10 kilograms. The most reliable method for delivering an ASD device across the atrial septum is with the use of both fluoresce and TEE guidance.28 The procedure is performed under general anes-

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Figure 4. Schematic representation of transcatheter atrial septal defect devices.

Figure 5. The Amplatzer septal occluder device.
thesis with endotracheal intubation. Recently, an alternative to TEE has been investigated. Intracardiac echocardiography (ICE), with the transducer positioned in the right heart, has the advantages of not requiring general anesthesia as well as providing superior images to TEE with the addition of full-colour flow and Doppler capabilities.29 Patients may be kept overnight after catheterization with repeat TEE performed before discharge.

Complications associated with transcather device closure are shown in Table 3. These include cerebral embolism leading to stroke, cardiac perforation, device embolization requiring urgent surgical intervention, and endocarditis.

### Table 3
**Reported Complications Associated with Transcatheter Device Closure**

- Problems with device centering
- Tachydysrhythmias
- Heart block
- Thickening of the atrial wall
- Cardiac or vascular perforation
- Vascular complications
- Stroke
- Device erosion
- Problems with device retrieval
- Structural fractures
- Thrombosis
- Device embolization
- Endocarditis

Few studies have been performed comparing surgery versus device closure and comparing different devices. A summary of outcomes of various transcather devices is shown in Table 4. Visconti et al30 investigated the neuropsychological outcomes after surgical versus interventional closure of secundum ASDs in children. After regression analysis for patient age and parent IQ, significantly lower full-scale and performance IQ scores were noted in patients in the surgical group, with no significant differences noted on other tests.30 A randomized trial will be necessary to establish whether the observed results represent deleterious effects of cardio-pulmonary bypass (required with surgical closure) or a methodological artifact. Since there have been few studies comparing surgery versus device closure, one study sought out to compare transcather device versus surgical closure of atrial septal defects using a clinical decision analysis.32 A decision tree was created based on a literature review which estimated the probabilities for each combination of possible outcomes following either surgical or device closure of a secundum ASD. Utilities for the various combinations of possible outcomes were obtained from standard gamble interviews of health care providers. The utilities were then multiplied by the probabilities and summed back to the decision node, in order to give a final utility score for surgery versus device closure. Despite an initial bias from those interviewed in favour of transcather device closure, systematic assessment of outcomes from the decision analysis favoured surgery. However, surgery was associated with some inherent disutility (ie. presence of a surgical scar, greater pain and recovery), and adjustment for this equalized the outcomes for the two procedures.

### Epilogue

While an ASD most commonly occurs as an isolated lesion, our patient had associated Ebstein’s malformation of the tricuspid valve. These patients often have a marked predisposition to atrial tachydysrhythmias, both due to right atrial distension from the malformation and associated tricuspid valve regurgitation, but also because of associated conduction bypass pathways and preexcitation, such as is seen with Wolf-Parkinson-White syndrome. While the presence of a bypass pathway was not evident on surface ECG in our patient, this suddenly became manifest with the development of a rapid supraventricular tachycardia, which resulted in poor cardiac output, cardiovascular collapse and sudden death.

Autopsy confirmed the findings of a grossly dilated right atrial chamber due to atrialization of the right ventricle proximal to the severely displaced and dysplastic tricuspid valve. The actual functional right ventricular cavity was very small. The tricuspid valve was thickened and poorly mobile, with adherence of the septal leaflet to the ventricular surface causing some obstruction to ventricular outflow, and poor coaptation of the leaflets which con-

### Table 4
**Comparison of Various Transcatheter Devices that are in Use to Close ASDs**

<table>
<thead>
<tr>
<th></th>
<th>Surgery</th>
<th>Amplatzer</th>
<th>Clamshell</th>
<th>Button</th>
<th>Angel-Wings</th>
</tr>
</thead>
<tbody>
<tr>
<td># Devices Implanted</td>
<td>&gt; 229</td>
<td>&gt; 400</td>
<td>&gt; 200</td>
<td>&gt; 50</td>
<td>&gt; 74%</td>
</tr>
<tr>
<td>Implantation success Rate</td>
<td>100%</td>
<td>94%</td>
<td>87%</td>
<td>7%</td>
<td>92%</td>
</tr>
<tr>
<td>Incidence of Severe Complications*</td>
<td>5-8%</td>
<td>1.7%</td>
<td>6%</td>
<td>7%</td>
<td>8%</td>
</tr>
<tr>
<td>% Complete Closure</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Immediate</td>
<td>52%</td>
<td>9%</td>
<td>20%</td>
<td>70%</td>
<td></td>
</tr>
<tr>
<td>3 months</td>
<td>95%</td>
<td>35%</td>
<td>35%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>6 months</td>
<td>47%</td>
<td>48%</td>
<td>48%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;12 months</td>
<td>55%</td>
<td>52%</td>
<td>52%</td>
<td></td>
<td></td>
</tr>
<tr>
<td>&gt;48 months</td>
<td>97%</td>
<td></td>
<td></td>
<td></td>
<td>74%</td>
</tr>
</tbody>
</table>

* = Severe complications include: death, cerebrovascular accidents, need for emergent operation, tamponade, cardiac arrest and severe dysrhythmias.
tibuted to the tricuspid regurgitation. The ASD closure device was seen (Figure 6) and appeared to be well endothelialized, although there were fractures of some of the wire struts. The device was non-mobile and adherent, and there were no communicating defects. Histologic examination showed the device to be covered with fibrous and endothelial tissue (Figure 7). In addition, sections of the atriop-ventricular ring revealed the presence of a muscular band consistent with a conduction bypass pathway.

Figure 6. Gross pathology section of the atrial septum. The ASD closure device appears to be well endothelialized except for the lower left corner. This edge is not well appositioned to the atrial septal wall and therefore was not endothelialized.

Figure 7. Histological slice of the ASD closure device. Dense fibrous tissue can be seen in the periphery (*) while areolar fibrous tissue is seen centrally (**). The fibers of the device are also seen (white arrow). Focal foreign body reaction (FBR) is noted in contact with the fabric of the device.

Acknowledgements
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References