Non-surgical Alternatives to Repair Congenital Heart Defects

a report by

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Children and adults with congenital heart disease are increasingly benefiting from non-surgical alternatives to repair their heart defects. The last decade has seen a remarkable advance in the tools and techniques used to treat congenital heart disease in the cardiac catheterization laboratory. This article summarizes a few of the important tools available in 2005 to treat some of these conditions.

Atrial Septal Defect

Secundum atrial septal defects (ASDs) are one of the most common congenital heart defects. The ASD is a hole in the atrial septum that allows a portion of blood returning to the left atrium to pass to the right atrium, right ventricle and pulmonary arteries, thereby placing additional work on these heart structures as well as the lungs. Left untreated, atrial septal defects may lead to right heart failure, atrial arrhythmias, ventricular dysfunction, and pulmonary hypertension.

Elective transcatheter closure of ASDs is currently indicated for patients with ASDs of the secundum type, with a weight of >15kg and evidence of right ventricular dilation. As a lot of ASDs are not diagnosed until adulthood, many are closed in the adult population.

Despite many attempts beginning in the 1970s to invent the ideal transcatheter ASD occluder, the first and only device to date to receive US Food and Drug Administration (FDA) approval for ASD closure is the Amplatzer Septal Occluder (ASO). This device was first approved in the US in 2002. The device is composed of a self-expanding nitinol wire frame that is introduced transvenously and passed across the atrial septum where the device is then placed in position across the defect (see Figure 1). Occlusive polyester fabric patches are sewn inside the frame. The device is collapsed into a delivery sheath (7–12 French) and is delivered through a 9 French sheath. One advantage of this device is its low-profile; disadvantages include the complexity of the delivery system, and inability to close defects larger than 22mm.

Patent Foramen Ovale

Poised on the cusp of FDA approval is the Helex Septal Occluder. The device design is a frame made of a single nitinol wire covered by an expanded polytetrafluorethylene (Gore-Tex®) membrane. In the heart, the device is formed into two round disks that sit on either side of the atrial septum. Because the device is not self-centering, the device disks must be significantly larger (the manufacturer recommends at least 1.6x) than the diameter of the defect to ensure that the entire defect is covered. The devices are available in disks ranging in size between 15mm and 35mm (in 5mm increments) and are delivered through a 9 French sheath. One advantage of this device is its low-profile; disadvantages include the complexity of the delivery system, and inability to close defects larger than 22mm.

While a patent foramen ovale (PFO) is found in ~15% of the normal adult population, it is found in over 50% of young adults with cryptogenic stroke. The implied mechanism is paradoxical embolization of clot from the right to left atrium and from there to the cerebral circulation. Because of the relatively low risk of recurrent stroke (1% to 5% per year), transcatheter closure of PFOs is currently indicated for patients with cryptogenic stroke and PFO who have had a recurrent stroke on medical therapy. There are two devices that currently have limited FDA approval—the Amplatzer PFO Occluder and the CardioSeal. The limited FDA approval (humanitarian device exemption (HDE)), while not investigational,

requires institutional review board oversight and does not permit ‘off-label’ usage. Randomized trials comparing medical therapy with device closure after a first cryptogenic stroke are under way.

PFO closure is also being investigated as a therapy for migraine headaches. Studies have shown a strong association between migraine headache and PFO. The proposed mechanism is paradoxical shunting of vasoactive substances to the cerebral circulation. Recent uncontrolled studies have shown a dramatic reduction in migraine frequency in those patients with PFO who have undergone transcatheter PFO closure. Several randomized controlled trials with a variety of transcatheter PFO occluders will start in the US by the end of the year.

**Patent Ductus Arteriosus**

The ductus arteriosus is a fetal blood vessel connecting the aorta with the pulmonary artery. Normally, this vessel closes in the first days of life; however, if it remains open, this may lead to pulmonary over-circulation, left atrial and ventricular dilation, and pulmonary hypertension. Even in the absence of a significant hemodynamic burden, a patent ductus arteriosus (PDA) is a nidus for endarteritis with a risk of ~1% per year. Indications for PDA closure are therefore any audible or hemodynamically significant PDA.

A variety of devices have been used ‘off-label’ to close PDAs. Small PDAs (<2mm) are easily closed with stainless-steel Gianturco coils. Larger PDAs are better closed with the Amplatzer Duct Occluder, which is a mushroom-shaped device with a nitinol frame and filled with an occlusive polyester fabric mesh. This device is delivered from the venous approach placing the ‘hat’ in the aortic ampulla and the ‘stem’ in the PDA itself (see Figures 3 and 4). Closure rates are virtually 100% for PDAs up to 10mm and complications are rare. Patients who are more than six months old or 6kg in weight are the best candidates for transcatheter PDA closure.

Due to the small delivery system (5–7 French), excellent occlusion characteristics, and low-profile, the Amplatzer PDA Occluder has also been used ‘off-label’ to occlude other congenital and acquired defects. Examples include closure of prosthetic paravalvular leaks, and residual patch-margin ventricular septal defects.

**Ventricular Septal Defect**

The most common ventricular septal defects (VSDs) are of the membranous type—located just underneath the aortic valve. There are currently no FDA-approved devices to close membranous VSDs. There is one membranous VSD occluder that is available elsewhere in the world. This device is eccentric with a recessed aortic margin to avoid interference with this valve. This

device is much more technically demanding to implant than the ASD or PDA devices. Reports of late complete heart block are worrisome and thought to be due to the constant radial force of the nitinol against the bundle of His, which is intimately associated with the posterior margin of the membranous VSD. Because the surgical results for membranous VSDs are generally excellent, any transcatheter device will need to meet or exceed this high standard to become an acceptable alternative.

Muscular VSDs are found elsewhere in the ventricular septum. While situated away from the conduction system, they are often multiple or associated with a membranous VSD. Unlike membranous VSDs, muscular VSDs, especially when multiple or located at the apex of the heart, are surgically challenging. The only transcatheter device that was FDA-approved to close these defects is the CardioSeal. This device is a double-umbrella device with four ‘arms’ of nitinol wire on each umbrella supporting a square Dacron patch. Although this device was originally designed to close atrial communications, the lack of surgical or transcatheter alternatives to address complex muscular VSDs allowed FDA approval of the CardioSeal device for muscular VSD closure. However, the large delivery system (11 French) and poor retrievability characteristics limit the utility of the CardioSeal device for this indication.

There is a muscular VSD occluder based on the same nitinol frame design as their ASD device, but with a thicker connecting waist and smaller retention disks to conform to the muscular septum. This device is much easier to retrieve and reposition, and has a smaller delivery system than the CardioSeal. It is in the final phases of FDA approval.

A VSD occluder specifically designed to treat post-myocardial infarction (MI) ventricular septal defects is, at present, limited to investigational use in the US.

**Stenotic and Regurgitant Lesions**

A mainstay of interventional congenital cardiology is the use of transcatheter balloon dilation and stenting to relieve vascular stenoses. Areas that frequently require dilation and/or stenting in the congenital population are the pulmonary arteries, aorta, and various conduits and venous baffles. Most of the balloons and stents used by congenital cardiologists were designed and approved for use in the peripheral vascular and biliary systems. In the past few years, several balloons have been specifically manufactured for congenital cardiology applications. One example is the ‘Balloon-in-Balloon’ (BiB), which is an innovative design with a smaller inner balloon and larger outer balloon. This design facilitates the delivery of large stents in structures, such as the pulmonary artery or aorta, limiting the risk of balloon rupture during stent placement.
The stents used by congenital cardiologists in the US are all adapted from other approved indications such as biliary and peripheral vascular stenting. Though designed for other applications, the availability of premounted stent-balloon combinations has greatly facilitated the delivery of medium-sized stents in small children (see Figure 5).

Early clinical trials are under way to evaluate the use of transcatheter delivery of stent-mounted valves. Many congenital heart patients have had reconstruction of the pathway from the right ventricle to the pulmonary artery with surgically placed, valved homografts. As the valve function of these homografts deteriorates over time, these patients can be left with significant pulmonary regurgitation—the deleterious consequences of which are increasingly being appreciated. Transcatheter placement of a new valve is desirable to avoid repeat surgery. The current technology is based on a bovine interval jugular valve mounted inside a balloon-expandable stent. Early work conducted in Europe developing this system appears promising. The long-term durability of these implanted valves remains to be seen.

Summary

Many tools to treat children and adults with congenital heart disease are devices adapted from other applications. Recently, devices specifically for use in congenital heart disease have been FDA-approved and have dramatically advanced the non-surgical care of patients with congenital heart disease. Further development of tools specifically designed for congenital heart applications are forthcoming and will continue to advance the field.