New Transcatheter Techniques for Creation or Enlargement of Atrial Septal Defects in Infants With Complex Congenital Heart Disease

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Objectives: To describe a series of 8 consecutive infants (5 with transposition of the great arteries [TGA] and 3 with hypoplastic left heart syndrome [HLHS]) who underwent nonconventional septostomy techniques. Background: For some complex congenital heart defects, an unrestrictive atrial septal defect (ASD) is essential to achieve an adequate cardiac output and/or systemic saturation. In some scenarios, the use of conventional septostomy techniques may be technically difficult, hazardous, and/or ineffective. Methods: Use of transhepatic approach, cutting balloons, and radiofrequency perforation with stenting of the atrial septum. Results: The size of the ASD and the oxygen saturation increased in all patients with no major complications. In those with TGA, the ASDs were considered to be of good size at the arterial switch operation. Two of the 3 patients with hybrid palliation for HLHS have developed some degree of obstruction within the interatrial stent over 2–3 months. At surgery, the stents were found to be secured within the septum with one showing significant fibrous ingrowth after uneventful removal. The other had some nonobstructive ingrowth. Conclusions: Creation or enlargement of ASDs in infants using new nonconventional transcatheter techniques is feasible, safe, and effective, at least in the short-to-mid-term follow-up. Infants with TGA seem to benefit the most because the procedure results in satisfactory clinical stability for subsequent early surgical intervention. In infants with HLHS palliated by a hybrid approach, stent implantation to the atrial septum seems to buy enough time to bring them to the phase II safely despite progressive in-stent obstruction.

Key words: atrial septal defects; congenital heart disease; atrial septostomy; stents; cutting balloons

INTRODUCTION

In some infants with complex congenital heart disease, an unrestrictive atrial septal defect (ASD) is required for adequate blood mixing at the atrial level or relief of right or left atrial hypertension resulting in better cardiac output and/or systemic saturation. Transcatheter creation or enlargement of an ASD is usually achieved employing a variety of techniques including transseptal puncture (if the septum is intact), balloon and/or blade septostomy, and static balloon dilation of the interatrial septum (IAS) [1–3]. However, due to the increased thickness of the IAS in infants beyond the neonatal period and in neonates with hypoplastic
left heart syndrome (HLHS), the use of balloon septostomy and/or static balloon dilation may not be effective to attain an adequate sized orifice. Blade septostomy followed by balloon dilation of the IAS has been described as an alternative approach in such situations [4,5]. However, the application of this technique may be associated with some technical limitations particularly in small babies, in whom the use of larger sheaths (required to accommodate the blade) may result in vascular damage, and in patients with a diminutive left atrium (LA), in whom the use of the blade may increase the risk of cardiac laceration [6]. Additionally, some occasional anatomical variants such as obstruction of the femoral veins or congenital interruption of inferior vena cava (IVC) can make the access to the IAS more difficult.

Recently, to circumvent the aforementioned problems, we have used a variety of new transcatheter techniques for creation and/or enlargement of ASDs. Although most (if not all) of them have been already described [4], their use has been confined to a limited number of patients and centers. Therefore, more information is needed to assess their reproducibility in different hands, safety, and efficacy. In this article we report the outcomes after the application of these techniques in a series of 8 consecutive infants with complex congenital heart disease with restrictive or absent ASDs.

PATIENTS AND METHODS

Study Design and Patient Population

This was a retrospective study in which data were collected through chart review. From January 2003 to December 2006, 8 consecutive infants with complex congenital heart disease and restrictive (7 patients) or absent ASDs (1 patient) underwent interventional procedures for creation or enlargement of the interatrial communications using a variety of techniques as described later. In these patients, classic balloon septostomy could not be technically performed, was initially ineffective or was not even contemplated due to an anticipated limited efficacy (too thick an atrial septum). The median age of the patients was 30 days (range: 3–102 days) and the mean weight was 3.3 ± 0.6 kgs (Table I). Patients were classified in two groups according to their main diagnosis: Group A: 5 patients with complete transposition of the great arteries (TGA); and Group B: 3 patients with HLHS. Of the 5 patients in group A, one had an intact IAS and 4 had a very thick IAS. The three infants in group B had been initially palliated without the use of cardiopulmonary bypass employing a variety of procedures including banding of the pulmonary arteries (all 3 patients) ± stenting of the ductus arteriosus (1
placement of a reversed modified (Goretex) shunt from the main pulmonary artery to the innominate artery (2 patients) ± placement of an 8-mm modified (Goretex) shunt from the main pulmonary artery to the descending aorta (1 patient). Two of these patients had had classic balloon septostomy and static balloon dilation at the time of the initial palliative surgery. All of them had developed a restrictive ASD within days to 3 months after the initial surgical palliative procedure.

Cardiac Catheterization

All patients underwent the interventional procedure in the catheterization laboratory under general endotracheal anesthesia. Informed consent was obtained from parents. Vascular access was established in the femoral vein in all but one patient. A transhepatic access was also needed in one patient with TGA. Heparin sulfate (100 units/kg) was administered after a 6 or 7 Fr sheath was placed in the femoral vein or through the liver. In those patients who required mechanical or radio frequency (RF) transseptal access, heparin was given only after the LA was entered. To delineate the plane of the IAS, angiograms were performed in postero-anterior and hepatoclavicular views using angiographic catheters or through the side arm of the sheaths. Prophylactic antibiotics were given for all patients in group B. Small blade septostomy catheters, such as the PBS 100 and 200 Park (Cook, Bloomington, IN), were not commercially available in the country in which these procedures were performed.

Imaging Monitoring

In addition to the traditional fluoroscopic guidance, the procedures were also monitored by transthoracic color Doppler echocardiography (TTE) in 3 patients from group A (Fig. 1A) and in one in group B (Table I). An AcuNav 5.5–10 MHZ phased-array single-plane intracardiac probe (Acuson/Siemens Mountain View, CA) connected to a Cypress machine (Acuson, Siemens) was used for transesophageal echocardiographic (TEE) monitoring in 2 patients from group B (Fig. 2A) and in 1 from group A (Table I).

Transseptal Puncture Techniques

In a 3 month-old infant with TGA, intact IAS and a restrictive VSD, transseptal access was mechanically obtained using a Brockenbrough transseptal needle advanced through a 6 Fr Mullins transseptal sheath/dilator set (Benson, Cook) and standard techniques [7]. The IAS was marked with some contrast material injected through the needle before its progression towards the LA. In this patient, the procedure was also monitored by TTE guidance (Fig. 1A). After the sheath was advanced towards the LA, a 0.014” angioplasty guide wire was left in the left upper pulmonary vein for subsequent balloon dilation of the IAS using cutting and standard catheter balloons (Fig. 1B).

Fig. 1. A: Transeptal puncture under echo guidance. A small injection of contrast material through the transeptal needle is seen after the left atrium was entered. B: The cutting balloon is inflated across the interatrial septum over a 0.014” guide wire and moved forward and backwards gently.
In two patients from group B, the restrictive ASD was located in the upper portion of the IAS near the superior vena cava (Fig. 2A). In both patients, a 5-Fr right coronary Judkins guiding catheter was pushed against the mid-portion of the IAS, below the native ASD. Small amounts of contrast material were injected to tag the IAS. Transseptal access was performed using a Nykanen RF perforation wire and a coaxial catheter (Baylis Medical Company, Montreal, Canada), as described in previously published protocols [8–10] (Fig. 2 B). In both cases, the IAS was perforated using an energy of 8 W for 5 sec. After the transseptal access was established, the coaxial catheter was advanced to the LA and the RF wire was exchanged for a 0.014" coronary wire. Over this wire the IAS was subsequently dilated using a 2.5 × 15 mm coronary angioplasty balloon. This allowed further wire and catheter exchanges and progression of a 6 Fr, 23 cm long sheath (Cordis) towards the LA. Through this sheath, a premounted Genesis stents (Cordis) was implanted across the IAS (Fig. 3A and B). In the other patient from group B (who was the first to undergo this kind of procedure), the stent was implanted across a native ASD located in the mid-portion of the IAS without the use of a long sheath.

Transhepatic Venous Access

This access was required in one neonate in group A with bilateral occlusion of the femoral veins because of previous catheter insertions in other centers. Vascular access via the umbilical vein was not possible due to vessel necrosis. As described in previous publications [11,12], a 22-gauge needle with a mandril was used to puncture the liver on the axillary midline, midway between the diaphragm and the lower edge of the liver. The needle was advanced in the direction of the spine, the mandril was removed, and repeat small injections of contrast medium within the hepatic parenchyma were performed until free flow through a hepatic vein towards the IVC and right atrium was observed. A regular 7-Fr sheath was placed through the liver over a guide wire. This allowed subsequent static balloon dilation of the IAS with cutting and regular catheter balloons followed by standard atrial septostomy. After completion of the procedure the intrahepatic route was occluded with two 38-5-5 Gianturco coils.

Cutting Balloon Septostomy

In all patients in group A, the atrial septostomy was initially performed using cutting balloons (Boston Scientific, IVT, San Diego, CA) ranging from 6 to 8 mm in diameter and with 10 mm in length. The cutting balloon catheter was advanced through a 6- or 7-Fr short sheath over a 0.014" coronary angioplasty wire or a 0.018" guide wire (Road Runner, Cook) positioned in the left upper pulmonary vein or curled in the body of the LA. The balloon was centered...
across the IAS (Fig. 1B) and inflated up to its nominal pressure. When inflated, it was moved slightly forward and backwards to enhance the incising effects of the microblades. It was slowly deflated and rotated in clock or counterclockwise direction followed by 2–3 new inflations with the hope to produce several tears around the margins of the ASD. This was followed by static balloon dilation using high pressure balloons.

**Static Balloon Septostomy**

This modality was performed in all patients in group A following cutting balloon septoplasty. After a standard 0.035", 260 cm long guide wire was positioned in the left upper pulmonary vein, a high pressure catheter balloon (Powerflex, Cordis) was inflated 2–4 times across the IAS. Maximum balloon diameter was 12 mm and the maximum inflation pressure was 12 atm.

**Stent Atrioseptoplasty**

All patients with HLHS from group B had stent implantation within the IAS over extra-support 0.014" coronary angioplasty guide wires using similar techniques reported in previous protocols [13]. In all procedures, a premounted Genesis stent (Cordis) ranging from 7 to 8 mm in diameter and from 19 to 26 mm in length was used. Stent diameters were chosen empirically as such to provide an unrestrictive and potentially durable (for at least 2–4 months) flow through the IAS. Stent lengths were also chosen arbitrarily as such to allow adequate stabilization within the IAS, minimizing the risks of embolization due to some movement during balloon inflation/stent deployment and/or due to foreshortening after its expansion. In the first patient, the stent was implanted across a native ASD without the use of a long sheath. In the others, it was implanted within a newly created ASD using RF technology and a long sheath. Stents were deployed using a slightly higher pressure than the nominal pressure of the balloon. In all patients, the procedure was guided by either TTE or TEE. No attempts at crossing the newly implanted stent with a catheter were made. Gradients across the IAS were assessed using Doppler technology through TTE or TEE.

**Follow-up**

All patients were followed clinically and echocardiographically and were referred for subsequent surgical procedures at their cardiologist’s discretion. After stent implantation within the IAS, the patients received aspirin (~5 mg/kg/day) to prevent thrombus formation. Data from the surgical reports were also recorded and used to assess outcomes.
Statistical Analysis

Basic statistical analysis was performed using a SigmaStat\textsuperscript{®} 2.0 for Windows (Jandel) software. quantitative data are presented as means with standard deviation or median with ranges as applicable, and were compared using a paired t test. The level of significance was set at 0.05.

RESULTS

All interventions were technically successful and completed without any complications. The size of the ASD increased in all patients from a mean of 1.8 ± 0.8 to 5.3 ± 1.1 mm (P < 0.001) and the arterial oxygen saturation measured by percutaneous oximetry improved from a mean of 63.9 ± 4.7 to 78.4 ± 4.2% (P < 0.001) (Table I). The mean pressure gradient across the IAS measured by catheterization in the 5 patients from group A decreased from a mean of 4.2 ± 1.0 to 0.6 ± 0.5 mm Hg. In one patient from group A, there was some difficulty pulling an 8 mm cutting balloon inside a 7 Fr sheath. This was solved by new inflations and slow deflations of the balloon. In the 3 patients from group B the mean pressure gradient estimated by Doppler echocardiography across the IAS by echocardiography decreased from to 18.0 ± 5.2 to 1.7 ± 0.6 mm Hg (P < 0.001). In these patients, the Doppler pattern in the pulmonary veins improved after stent implantation, with a less prominent retrograde A wave. In one patient from group B, in whom the stent was implanted through the native ASD, the implant remained in an off-center, albeit stable, position with about 2/3 of its length occupying the right atrium. No impingement on the tricuspid valve was observed on TTE. No hemodynamic compromise, airway, or esophageal complications were observed after the use of the AcuNav probe (Siemens/Acuson) for TEE monitoring. All, but one patient, were extubated within 24–48 hr and remained clinically well. On follow-up, the 5 patients from group A underwent surgical repair (arterial switch operation) 3–15 days after catheterization with one surgical death. At surgery, the ASDs were considered to be of good size. Two of the 3 patients from group B (patient 6 and 7) have developed some degree of obstruction within the interatrial stent over 2 months, as determined by TTE. In these patients the transatrial gradient increased from 2 mm Hg at the time of the procedure to 20 mm Hg (patient 6) and to 12 mm Hg (patient 7) at the last assessment. Cardiac catheterization performed in patient 7 before the comprehensive phase II operation showed a distal mean pulmonary artery pressure of 17 mm Hg, with a mean wedge pressure of 11 mm Hg and a right atrial pressure of 9 mm Hg. Both patients have undergone a bidirectional cavopulmonary anastomosis along with debanding of the pulmonary arteries, reconstruction of the neoaorta, and atrial septectomy with one surgical death (patient 7). At surgery, the stents were found to be appropriately secured within the IAS with one of them remaining in an off-center position, as diagnosed before (patient 6). This stent also showed significant fibrous ingrowth after uneventful removal. No thrombus formation was observed within the stent struts. The other stent (patient 7) had some ingrowth, albeit not obstructive. The remaining patient from group B (patient 8) is awaiting further surgical management with no significant gradient across the IAS after a month following IAS stent implantation.

DISCUSSION

Standard balloon atrial septostomy has become one of the major indications for interventional cardiac catheterization in the neonatal period [1–2]. In some challenging cases, blade septostomy and static balloon dilatation may be used as adjunctive modalities to improve outcomes [4–6]. However, there are some isolated or combined situations in which these techniques may be technically impossible, difficult, risky, or ineffective such as a thick or intact septum, a small LA, a small baby, occluded femoral veins, absence of the hepatic portion of the IVC, or unavailability of an adequate size Park blade. In this article we have described some technical alternatives to overcome some of these limitations, achieving satisfactory short-term outcomes.

In newborns with an intact (and invariably thick) IAS the first challenge is to gain access to the LA. Since its original description by Ross et al. [14], the transseptal puncture technique has undergone several modifications allowing its application even in small infants with safety and efficacy [7–9], as seen in one patient in this series. Mechanical perforation of the IAS with the Brockenborough needle may not be feasible or safe when there is no direct approach to the IAS or capability for the needle to be forcefully impinged on and pushed against the IAS. Examples of these situations include venous access through the jugular vein, no enlargement of the LA, and no bulging of the IAS into the right atrium [7]. Also, it has been suggested that when the IAS is too thick and the LA is small, a combination commonly observed in neonates with HLHS, the use of the long transseptal needle may carry a higher risk of inadvertent cardiac perforation [15]. Radiofrequency assisted perforation of the IAS, as seen in 2 patients in this series, has been proposed as a technical alternative to these patients [10,15,16]. The main advantages of the RF system for transseptal access include the possibility of perforating...
even an unusually oriented IAS through the inferior or superior vena cava approach using preformed, softer guiding catheters; and elimination of the potentially dangerous strong forward force necessary to push the needle through a thick IAS. Because the RF wire delivers the RF energy to a very tiny spot producing a minute hole, even inadvertent perforations in undesirable locations may be well tolerated as long as the coaxial catheter is not advanced over the RF wire. As such, it seems that the use of the RF assisted transseptal access in infants, especially those with unfavorable underlying septal anatomies, results in a much more controlled, smoother perforation, which may help to decrease the complications associated with the mechanical transseptal needle puncture.

In this series, both types of transseptal puncture were monitored by fluoroscopy and echocardiography. Although this latter adjunctive imaging modality is not mandatory for a safe access to the LA [4], the authors felt that in the patient with TGA who underwent mechanical perforation, the TTE was helpful to confirm a “give” as the tip of the needle popped into the LA. This was especially important to prevent perforating the roof of the LA considering the strong forward force that was required to push the transseptal needle through the thick IAS. Moreover, the use of echocardiography in most of our cases helped to expedite the assessment of the size and adequacy of the interatrial communications, limiting further catheter manipulations, and angiograms, which is important in these critically ill infants. As previously described in a patient with HLHS and intact IAS [16], we have also employed the 10 Fr AcuNav (Siemens/Acuson) probe for TEE monitoring during RF transseptal perforation. In small infants, the use of the higher profile biplane or multiplane pediatric TEE probes may cause airway and, especially, LA compression, which may distort the underlying septal anatomy and limit even further the already restricted space in the LA for stent deployment. Although the AcuNav (Siemens) is a monoplane probe, the quality of the pictures was considered to be satisfactory in our cases in which it was employed. Although the AcuNav does not have an attached thermistor, thermal damage in the esophagus does not seem to be an issue [16].

Occlusion of the femoral veins, found in a case in this series, has been considered another anatomical limitation for a standard balloon septostomy. Although the umbilical vein is a good alternative in these cases, this vessel was obstructed at the time of the procedure. The transhepatic access has been used for a variety of procedures in the cardiac catheterization laboratory [11,12]. The advantages of this route include a better angle to access the atrial septum and the possibility of using larger sheaths in small children without vascular damage. Its feasibility and safety has been demonstrated in infants and children [11,12,17]. Although complications are not frequent, occurring in less than 5% of cases, intraperitoneal or intrapleural bleeding, pulmonary embolism, and peritonitis may all be encountered [18].

After reaching the LA, the increased thickness of the IAS may be another limiting factor for conventional septostomy, decreasing its efficacy and theoretically increasing the risks of avulsion of the pulmonary veins and the LA [1–3]. To overcome this limitation, static high pressure balloon dilation of the IAS has been used successfully to enlarge ASDs in infants [19]. However, these communications are often not durable and manifest increased stenosis over time [15,19]. Although there is limited data on the role of cutting balloons to achieve long lasting ASDs, short-term results are encouraging [15,16,20,21]. Hill et al. [16] have suggested that the microsurgical blades of the cutting balloon allows controlled tearing of the septal wall rather than stretching of the thickened IAS, as seen with static balloon dilation. We propose that rotation of the balloon followed by repeat inflations may tear the IAS in different locations and improve the response to static balloon septoplasty. All newborns in group A in our series have undergone this kind of septostomy with marked improvement of the clinical conditions and production of adequate sized ASDs, confirmed at surgery.

The issue of which is the best method to provide an unobstructed and long lasting flow through the IAS in patients with HLHS undergoing initial palliation without cardiopulmonary bypass remains to be determined with ongoing experience. Galantowicz and Cheatham addressed the technical difficulties related to enlargement of ASDs in this difficult group of patients [15]. Standard balloon septostomy seems to be less effective and even hazardous because of the thickened IAS, small LA, and the posteriorly placed ASD. Blade septostomy also has several limitations and hazards because of the smallish size of the LA. Static balloon septostomy, even preceded by cutting balloon septostomy, may relieve the transatrial gradient initially but does not seem to promote a durable unrestrictive ASD. As such, these authors went on to recommend stenting the IAS when it is thick enough [15]. In one recent small series [22] atrial septal stent implantation has been shown to result in sustained hemodynamic benefit in infants with left sided obstructive lesions. Other reports also confirm the usefulness of stenting the IAS in different clinical scenarios [13,15,23]. With evolving stent technology, low profile, premounted, rapid exchange, and medium-sized stents have become
available for the interventionalist and constitute an attractive technical alternative to stent the IAS in such small and fragile patients. However, stent malposition with impingement on the superior or IVC, pulmonary veins, and tricuspid and mitral valves; stent embolization and possible thrombus formation upon the stent struts remain significant potential complications. Malposition of the stent was observed in one patient in this series without interfering in other cardiac structures. Although this patient was the first to undergo this type of procedure in this series, the authors feel that the use of a long sheath may minimize this complication by adding some additional support to maintain a forward force while the stent is being deployed. From the technical standpoint, it has not been clear whether stenting the IAS through a new RF created hole is better than stenting the native ASD. We think that creating a new hole in a different portion of the IAS offers more support for stent fixation, minimizing the risk of stent migration to the IVC, which could compromise the Fontan completion in the future.

The observation that 2 of our patients showed progressive obstruction through the interatrial stent, as determined by echocardiography, was of concern. However, it is possible that some overestimation occurs by using the mean Doppler gradient since one of our patients had satisfactory distal pulmonary artery pressures and a low mean gradient across the IAS at catheterization. On the other hand, the surgical findings of significant fibrous ingrowth within the explanted IAS stent in the other patient were disappointing. Whether this was related to the type of stent employed, its position (off-center) or final diameter is speculative. However, it may as well be that buying 2–5 months is all what is needed to get these patients to the comprehensive phase II operation.

This study has obvious limitations such as its retrospective nature and the small number of patients and procedures.

CONCLUSION

Creation or enlargement of ASDs in infants with complex congenital heart disease using a variety of transcatheter techniques including RF perforation, cutting balloons, transhepatic approach, and stent implantation is feasible, safe, and effective, at least in the short-to-mid-term follow-up. Infants with TGA seem to benefit the most because the procedure results in satisfactory clinical stability for subsequent early surgical intervention. In infants with HLHS and a restrictive ASD, stent implantation to the IAS seems to offer a longer lasting result when compared to other techniques, despite the finding of progressive stent ingrowth.

The issues of the ideal technique for stent implantation, ideal stent diameter and length, and progressive obstruction within the stent over time remain to be clarified with ongoing experience and larger number of patients.

REFERENCES