CONGENITAL CARDIOLOGY TODAY

Timely News and Information for BC/BE Congenital/Structural Cardiologists and Surgeons

Upcoming Medical Meetings

26th International Pediatric Association Congress of Pediatric Cardiology
Aug. 5-9; Johannesburg, South Africa
www2.kenes.com/ipa/Pages/Home.aspx

Cardiology Update 2010: The Heart of the Matter
Aug. 8-11, 2010; Whistler, Canada
www.mayo.edu/cme

ISHAC 2010
Aug. 31 - Sep. 2, 2010; Columbus, OH, USA
www.hybridsymposium.com

5th European Echocardiography Course for Heart DIsease
Oct. 6-9, 2010; London, UK
www.echocardiography-course.com

Transposition of the Great Arteries in the Neonate

By P. Syamasundar Rao, MD

INTRODUCTION

In the previous issues of *Neonatology Today (Neonatology Today is a sister publication to Congenital Cardiology Today)*, I discussed perinatal circulation,[1,2] an approach to the diagnosis of cyanotic neonate,[3] principles of management of the neonate with congenital heart disease[4] and neonatal cardiac emergencies[5] -- all addressing the general topics of congenital heart disease in the neonate. In this and future issues, commonly encountered cardiac defects in the neonate.

TRANSPOSITION OF THE GREAT ARTERIES

Transposition of the great arteries (TGA) is the most common cyanotic congenital heart defect (CHD) in the neonate. It constitutes 5% of all CHDs and 10% of all neonatal cyanotic CHDs.[6] A number of definitions have been used to describe TGA, but the most accurate description is "a defect in which the aorta arises from the morphologic right ventricle and the pulmonary artery from the morphologic left ventricle." In the most common form, referred to as complete transposition, the atria are normal in position (atrial situs solitus), there is atrio-ventricular concordance (right atrium connected to the right ventricle and the left atrium to the left ventricle), d loop of the ventricles (right ventricle is on the right and left ventricle on the left), ventriculo-arterial discordance (aorta arising from the right ventricle and the pulmonary artery from the left ventricle) and the aortic valve is located to the right of pulmonary valve (d-TGA). Thus the systemic venous blood from the vena cavae enters the right atrium and right ventricle and from there into the aorta while the pulmonary venous blood enters the left atrium and left
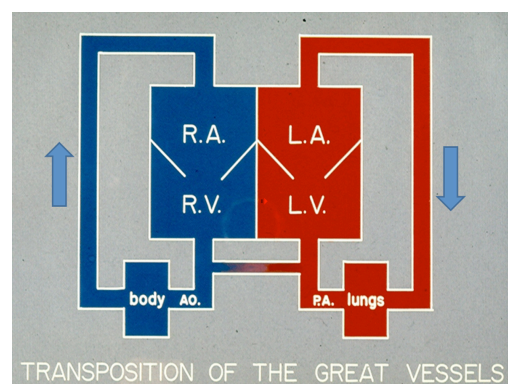

Figure 1. Box diagram of the heart showing parallel circulations in transposition of the great arteries. Note that right ventricle (R.V.) pumps into the aorta (Ao.) (because of transposition) which goes to the body and returns into right atrium (R.A.) and back into the body. Similarly left ventricular (L.V.) output goes to the pulmonary artery (PA.) and lungs and returns back to the left atrium (L.A.) and left ventricle to be pumped back into the lungs. Unless there are inter-circulatory communications via either a patent foramen ovale or patent ductus arteriosus, the infant cannot survive. Mixing across a ventricular septal defect (VSD) if such is present (not shown in the diagram) would also prevent progressive hypoxemia and death.

ventricle and from there into the pulmonary artery (Figure 1). Therefore, the circulation is parallel instead of normal in-series circulation. Therefore, the pulmonary venous blood does not get delivered to the body and the systemic venous blood does not get oxygenated. Infants will not live unless there are inter-circulatory connections such as atrial or ventricular septal defect or a patent ductus arteriosus.

CLASSIFICATION

The TGA patients are arbitrarily divided into: Group I, TGA with intact ventricular septum; Group II, TGA with ventricular septal defect (VSD), and Group III, TGA with VSD and pulmonary stenosis (PS).

CLINICAL FEATURES

Clinical features depend upon the anatomic type.

Symptoms

In Group I with intact septum, infants usually present with cyanosis within the first week of life (sometimes within hours to days of life). They may otherwise be asymptomatic. However, they will, with time, develop tachypnea and respiratory distress. If they are not appropriately treated, they become acidotic and go on to become lethargic without lack of spontaneous movement, and eventually die.

Group II TGA patients with VSD present with symptoms of congestive heart failure (tachypnea, tachycardia, sweating, and poor feeding) between 4 to 8 weeks of life, but the cyanosis is minimal.

Group III patients (TGA with VSD and PS) have variable presentation, depending upon the severity of PS and the degree of inter-circulatory mixing. If there is poor mixing, they may present early in life and mimic TGA with intact septum. If the PS is severe, the presentation is essentially similar to that seen with Tetralogy of Fallot (TOF).[7] With moderate PS the presentation is late with longer survival. With mild PS, congestive heart failure signs may be present, similar to Group II patients.

Physical Examination

The Group I patients with intact septum usually have severe cyanosis, but are without distress until severe hypoxemia and acidosis develop. Clubbing is not present in the newborn period and may not develop until 3 to 6 months. The right ventricular impulse is increased and the second heart sound is single. No cardiac murmurs are present; occasionally a grade I-II/VI nonspecific ejection systolic murmur may be heard along the left sternal border.

In Group II patients, tachypnea, tachycardia, minimal cyanosis, hepatomegaly, increased right and left ventricular impulses, single second sound, a grade III-IV/VI holosystolic murmur at the left lower sternal border and a mid-diastolic flow rumble (murmur) at the apex may be present.

In Group III patients, the findings are similar to TGA with intact septum, TGA with VSD, or TOF depending upon the degree of
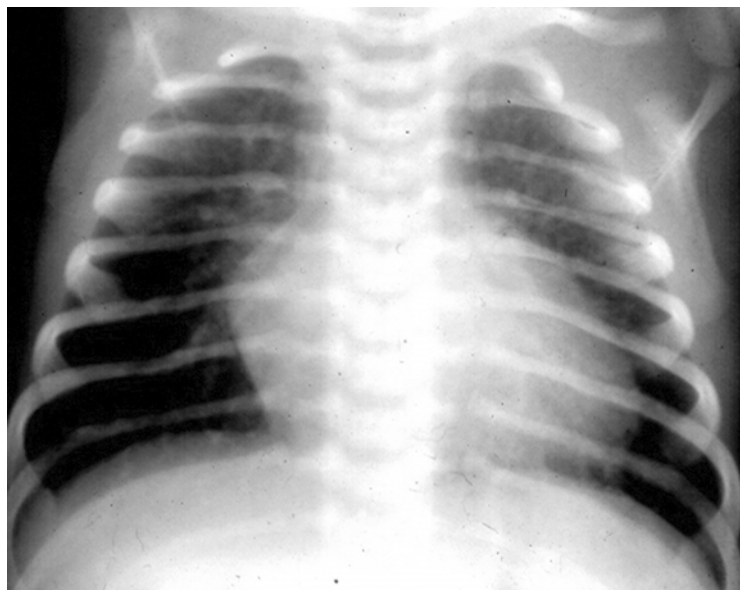

Figure 2. Chest radiograph of a two-day old infant with transposition of the great arteries demonstrating mildly enlarged size of the heart and increased pulmonary vascular markings.

mixing and severity of PS. Most of them however, will have a long ejection systolic murmur at the left upper sternal border and/or a holosystolic murmur at the left lower sternal border; both murmurs are usually grade III to IV/VI in intensity.

NONINVASIVE EVALUATION

Chest X-ray

In Group I patients with intact ventricular septum, chest roentgenogram looks benign with normal to minimal cardiomegaly and normal to slightly increased pulmonary vascular markings (Figure 2). The shadow of the thymus rapidly involutes and a narrow pedicle (superior mediastinum) may be seen. A combination of the above signs may sometimes appear as an "egg-shaped" heart on a postero-anterior chest film. In Group II patients with VSD, moderate to severe cardiomegaly and increased pulmonary vascular markings are usually seen. In Group III patients, mild to, at worst moderate cardiomegaly may be observed. The pulmonary vascular marking may be increased, normal or decreased, dependent upon the severity of PS.

Electrocardiogram

The electrocardiogram in a neonate with TGA and intact septum (Group I) may be normal with the usual right ventricular preponderance seen at this age. In older infants clear-cut right ventricular hypertrophy becomes obvious and, in addition, right atrial enlargement may be seen. In Group II patients, biventricular hypertrophy and left atrial enlargement are usual. In Group III, right ventricular or biventricular hypertrophy is seen.
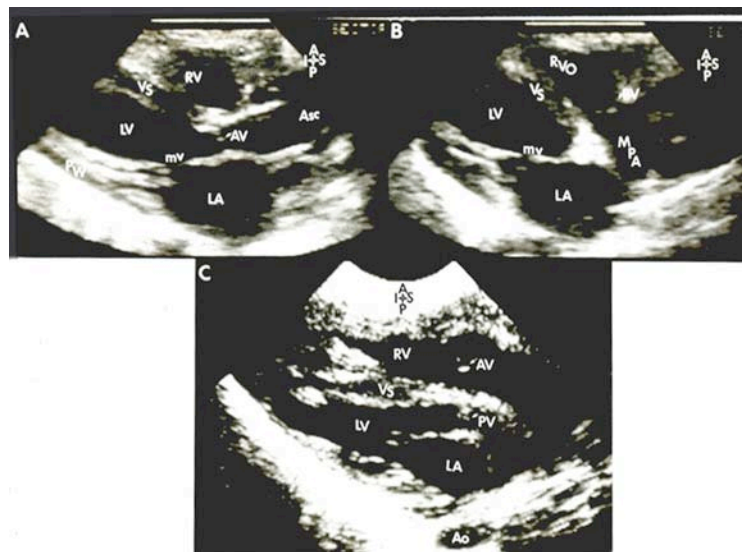
Figure 3. Precordial long axis echocardiographic views of two neonates: first (top, A & B) with normally related great arteries and the second (bottom, C) with transposition of the great arteries. In A, note that the posterior vessel arising from the left ventricle (LV) courses somewhat anteriorly, indicating that it is likely to be the aorta. In B, the anterior vessel coming off the right ventricle (RV) divides into right and left pulmonary arteries, suggesting that this vessel is main pulmonary artery (MPA). In C, the posterior vessel is coursing backward (posteriorly) after its origin from the LV and is likely to be the MPA, suggesting transposition of the great arteries. Ao, aorta; Asc, ascending aorta; AV, aortic valve; LA, left atrium; mv, mitral valve; PV, pulmonary valve; PW, posterior wall of LV; RVO, right ventricular outflow tract; VS, ventricular septum.

Echocardiogram

Echocardiogram is helpful in the diagnosis and assessment. Demonstration of transposition of the great arteries is somewhat difficult in view of the fact that atrial and ventricular anatomy is normal and the aortic and pulmonary valves appear similar on echocardiographic study. A helpful indirect sign is a somewhat posterior of the great vessel arising from the left ventricle in a precordial long axis view, indicating the vessel is pulmonary artery in contradistinction to anteriorly coursing ascending aorta (Figure 3). If one can follow the great vessel arising from the left ventricle and demonstrate its bifurcation (Figure 4), identifying it as a pulmonary artery, the diagnosis is easy. On-end visualization of both the aorta and pulmonary artery simultaneously on a precordial short axis view of the heart is also helpful in suggesting TGA. The presence of an interatrial communication and patent ductus arteriosus and shunt across them by color and pulsed Doppler should also be evaluated. In addition to these, demonstration of VSD and PS will place the patients into the respective groups.

Other Laboratory Studies

Blood gas values are useful in demonstrating the degree of hypoxemia and ventilatory status. Serum glucose (or Dexrostix), calcium,
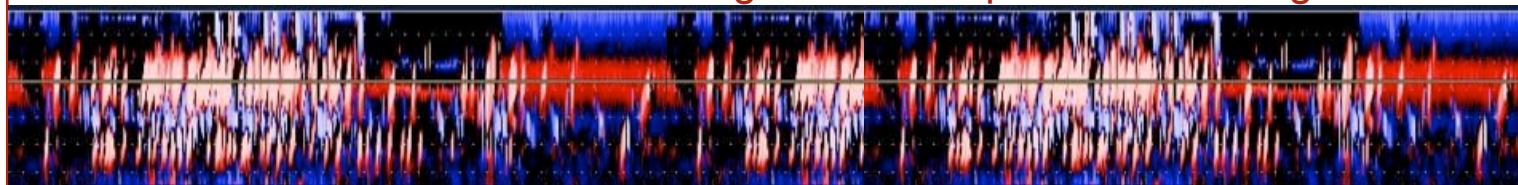
Figure 4. Selected video frames from a 2-dimensional echocardiographic view of an infant with transposition of the great arteries. In a, note the great vessel coming off of the left ventricle (LV) courses posteriorly and bifurcates into left (LPA) and right (RPA) pulmonary arteries. In b, posterior vessel is similarly seen to bifurcate. The anterior vessel is aorta (Ao). LA, left atrium; RV, right ventricle.
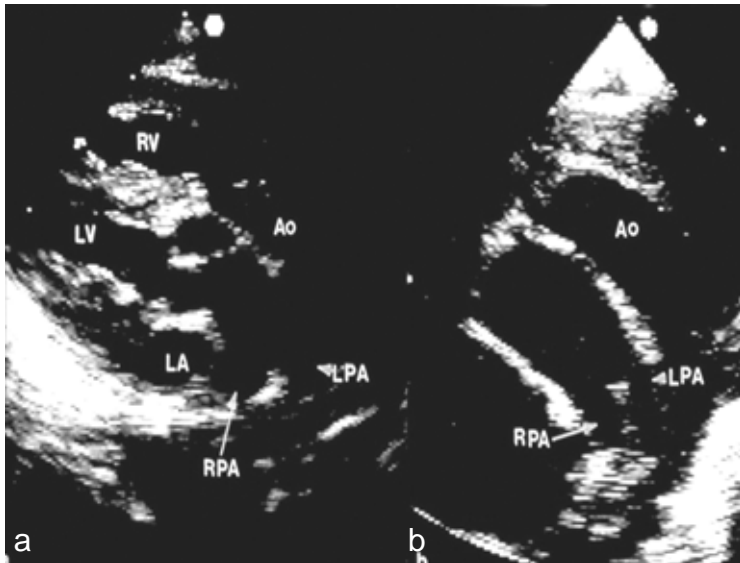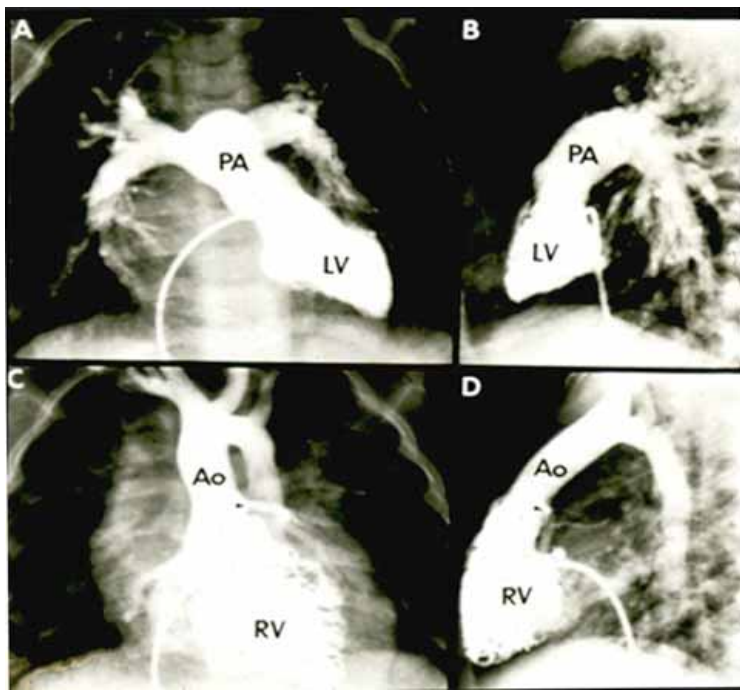

Figure 5. A & B. Left ventricular (LV) cineangiogram in postero-anterior (A) and lateral (B) views demonstrating a finely trabeculated, morphologic left ventricle with prompt opacification of the pulmonary artery (PA). The PA is inferior and posterior to its usual position. C & D. Right ventricular (RV) cineangiogram in postero-anterior (C) and lateral (D) views showing a coarsely trabeculated ventricle with opacification of the aorta (Ao). Note that the aortic valve is superior and anterior (D) to its usual position.

hemoglobin and hematocrit levels are useful in the overall assessment, similar to that of other cyanotic CHD in the neonate.[4]

CARDIAC CATHETERIZATION AND ANGIOGRAPHY

With the increased accuracy of echocardiographic diagnosis, invasive studies are not necessary for diagnosing TGA. Need for rapid relief hypoxemia and acidosis by balloon atrial septostomy and the need for a greater definition of coronary artery anatomy prior to arterial switch procedure may necessitate catheterization and angiography.

In Group I patients, vena caval, right atrial, right ventricular and aortic saturations are moderate to severely diminished unless atrial, ventricular or ductal shunting is present. Similarly, the pulmonary venous, left atrial, left ventricular and pulmonary arterial saturations are high with minimal, if any right-to-left shunt. In TGA, the pulmonary artery saturations are higher than those in the aorta; this is in contradistinction to higher aortic saturation in normal babies.

The left atrial pressure is usually high with a pressure gradient across the atrial septum. The right ventricular pressure is at systemic level without any gradient across the aortic valve. In TGA with intact septum, the left ventricular and pulmonary artery pressures are normal without any gradient across the pulmonary valve. However, in the early neonatal period, prior to involution of the pulmonary vasculature, these pressures are elevated, compared to normal right (pulmonary) ventricular pressure. In the presence of significant VSD and/or PS, the left ventricular pressure is elevated and this is usually proportional to the size of the VSD and/or severity of the PS. The pulmonary artery pressure is usually increased with associated VSD while with PS, it may be low to normal.

Selective right ventricular angiography (Figure 5) reveals a morphologically right ventricle with opacification of an anteriorly and superiorly displaced aorta. The aortic valve is located to the right of the pulmonary valve (d-TGA). The aorta ascends in a normal fashion and usually descends on the left side of the spine. The size and function of the right ventricle and presence of tricuspid insufficiency should be evaluated. If a VSD is present, it may be visualized. A laid-back view of the aortic root angiography along with a lateral view may be useful in demonstrating coronary artery anatomy. Aortography may, in addition, be useful in demonstrating PDA and coarctation of the aorta. Left ventricular cineangiogram (Figure 5) reveals a morphologic left ventricle with prompt opacification of the pulmonary artery. The pulmonary valve is located posterior, inferior and to the left of the aortic valve. Left ventricular angiography should be scrutinized for subvalvar and valvar PS. A VSD may be visualized, if present.

MANAGEMENT

The treatment of choice in the neonates with TGA is total surgical correction by arterial switch procedure (Jatene)[10] and will be discussed here-under. However, since the surgery is usually performed at about the age of 7 days, the infant should be cared for to ensure good clinical and metabolic state before going to surgery.

General Measures

Initial management of TGA is similar to that used in other cyanotic neonates.[4] The infant's temperature should be monitored and neutral thermal environment maintained. Ambient oxygen should be administered if the infant is hypoxemic. In cyanotic CHD patients, no more than 0.4 FIO_2 is necessary; higher levels of O_2 do not increase O_2 saturation because of fixed intra-cardiac right-to-left shunting. Metabolic acidosis, defined as pH <7.25 should be corrected with sodium bicarbonate (usually 1-2 mEq/kg diluted half and half with 5% or 10% dextrose solution) immediately. In the presence of respiratory acidosis, appropriate suctioning, intubation and assisted ventilation should be undertaken. Since hypoglycemia can be a significant problem, the infant's serum glucose should be monitored. The neonates should routinely receive 10% dextrose in water intravenously. If hypoglycemia (<30 mg/100ml) is detected, 15% to 20% dextrose solution should be

infused. Calcium levels should also be monitored and if hypocalcemia is detected, it should be treated.

Palliative Therapy

If untreated, *TGA with intact septum* carries a poor prognosis. Instead of having a normal in-series circulation, the TGA patients have parallel circulation (Figure 1). Without either an intra-cardiac or extra-cardiac shunt, the infants with TGA will not survive (Figure 6). The fetal circulatory pathways {patent foramen ovale (PFO) and patent ductus arteriosus (PDA)} will provide some mixing initially. However, in most neonates with TGA, the PFO and PDA tend to undergo spontaneous closure and the infant gets progressively hypoxemic. The PDA and PFO can be kept open/enlarged by pharmacological or mechanical means, respectively.

Patent Ductus Arteriosus. Intravenous infusion of prostaglandin E_1 (PGE_1) (0.05 to 0.1 mcg/kg/min) may help open the ductus, thus improve oxygenation. A small ductus may be made to dilate with PGE_1, but an already closed ductus may be difficult to reopen. Side effects include apnea, hyperthermia, muscular twitching and flushing. The side effects have not posed substantial management problems, but the neonate should be watched closely for apnea. Once the O_2 saturation improves, the dosage of the PGE_1 should be reduced stepwise downward to 0.02 to 0.025 mcg/kg/min. This may avoid the need for endotracheal ventilation because of apnea. If hypoxemia does not improve even after PGE_1, balloon atrial septostomy may become necessary.

Patent Foramen Ovale. Balloon atrial septostomy[8,9] (Figure 7) has been extensively used in the palliation of neonates with TGA with intact septum. The past experience has demonstrated that the improved mixing at the atrial level allows the neonate with transposition to grow to an age (usually 3 to 6 months) at which time a venous switch (Mustard or Senning - see surgical correction) procedure could safely be performed. With the introduction of arterial switch (Jatene) procedure

which is usually performed at approximately one week of age, balloon atrial septostomy is not necessary in all babies. If naturally present PFO and/or PGE_1 infusion to dilate the PDA do not maintain reasonably good oxygen saturations (60 to 70% without metabolic acidosis), balloon atrial septostomy should be performed, preparatory to arterial switch procedure.

Septostomy Procedures. In 1966, Rashkind and Miller[8] described a technique, now called Rashkind balloon atrial septostomy, which was extensively used to improve atrial mixing in neonates with TGA. It was subsequently applied to many other disease entities (reviewed elsewhere[11]) in which enlarging the atrial defect is beneficial. The reason for success of balloon septostomy is a very thin and frail lower margin of the PFO (septum primum) in the newborn which can be torn by rapid withdrawal of an inflated balloon across the PFO. Some babies do have thick atria septae. To address these situations, Park and his associates, in the mid/late 1970s, extended the utility of the balloon septostomy procedure by introducing blade atrial septostomy to enlarge defects with thick atrial septae.[12] A built-in retractable blade (knife) cuts the lower margin of the patent foramen ovale (PFO) which is followed by balloon atrial septostomy. More recently, static balloon angioplasty,[11,13,14] stents,[15-17] Brockenbrough atrial septal puncture,[17] radiofrequency ablation[18-20] and cutting balloons[20] were applied to create and/or enlarge the atrial defects.[9] In most patients conventional balloon atrial septostomy is all that is necessary to palliate TGA patients until surgery. In over a 35-year experience of the author, there were only a couple of occasions when static balloon[11,14] was used in TGA neonates. Other methods of atrial septostomy[9,15-20] are not necessary in TGA patients, but may be needed in patients with Hypoplastic Left Heart Syndrome.

Rashkind Balloon Atrial Septostomy Procedure. In TGA patients who are stable, the hemodynamic (usually limited) data, including selective cine-angiography, as needed, are performed. If the infant is unstable or has extremely low oxygen saturations, one may proceed directly with balloon septostomy. In such situations, aortic saturation and pressure pullback across the atria and echocardiographic size of atrial defect are
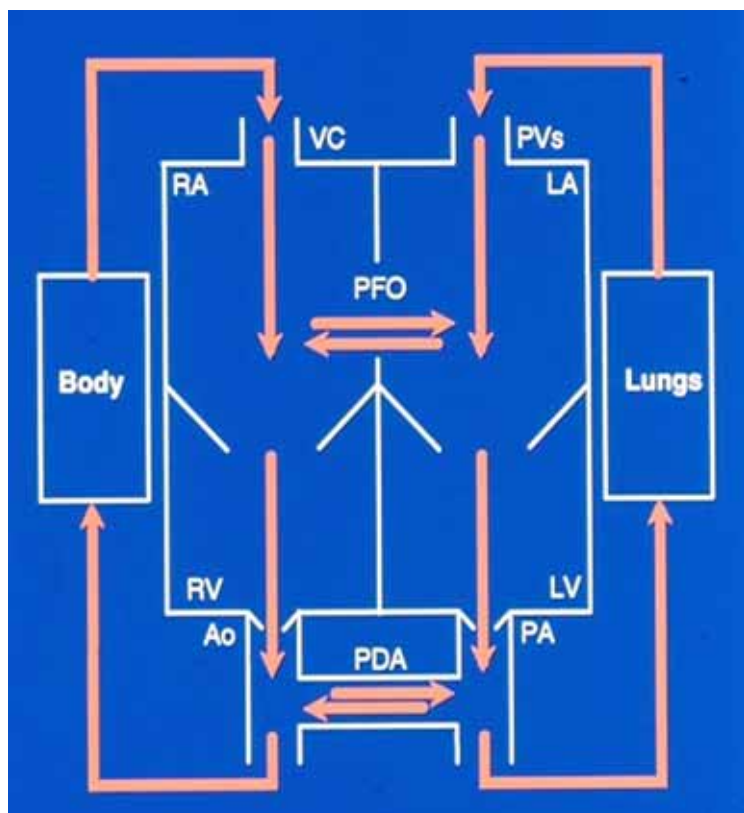
Figure 6. Box diagram of the heart showing parallel circulations in transposition of the great arteries. Unless there are inter-circulatory communications via either a patent foramen ovale (PFO) or patent ductus arteriosus (PDA), the infant cannot survive. Mixing across a ventricular septal defect, if such is present (not shown in the diagram), would also prevent progressive hypoxemia and death. Ao, aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; PVs, pulmonary veins; RA, right atrium; RV, right ventricle; VC, vena cavae.
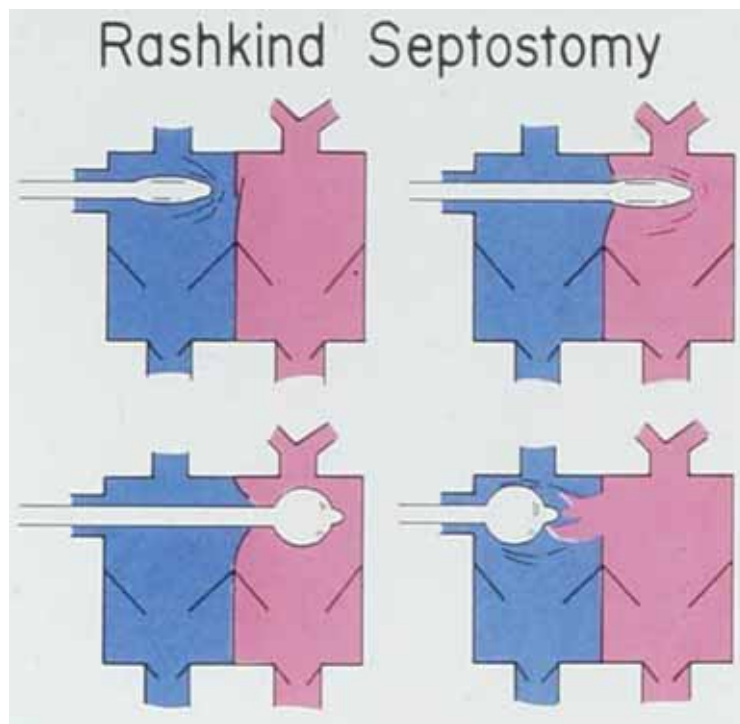

Figure 7. Diagrammatic display of the procedure of Rashkind balloon atrial septostomy. An un-inflated balloon septostomy catheter is placed in the right atrium (top left) and advanced across the patent foramen ovale into the left atrium (top right). The balloon is inflated with diluted contrast material (bottom left) and rapidly pulled back into the right atrium (bottom right), thus performing balloon atrial septostomy.

recorded. The balloon septostomy procedure involves inserting a balloon septostomy catheter, usually via a sheath percutaneously placed in the femoral vein, into the left atrium via the PFO. The balloon is inflated with diluted contrast material to a sub-maximal amount (usually 2 to 3 ml) and rapidly pulled back across the atrial septum (Figure 8) after ensuring that the catheter tip is located in the left atrium either by lateral fluoroscopy or by echocardiography. Once the catheter is pulled back to the inferior vena cava, the catheter should be rapidly advanced into the right atrium; all this is done as a single motion (Figure 8). The balloon should be deflated as the catheter is repositioned into the right atrium. This jerking motion of the contrast filled balloon catheter produces a tear in the lower margin of the PFO (septum primum) with resultant bidirectional shunt (Figure 9). We usually perform one additional septostomy following what may be considered good septostomy.

Increase in systemic arterial oxygen saturation, disappearance of pressure gradient across the atrial septum and echographic increase in the size of the atrial defect with non-restrictive Doppler flow across the

atrial septum are demonstrated in successful procedures. Some workers balloon-size the atrial defect both prior to and following balloon septostomy, and this is another method of assessment of the result of the septostomy.

In the initial description of balloon septostomy by Rashkind and Miller,[8] the catheter was introduced into the femoral vein by cut-down. To avoid femoral venous cut-down, insertion of the catheter and performance of balloon septostomy via the umbilical vein[21] has been advocated. When percutaneous technology became available, the balloon catheter was introduced via appropriate sized percutaneously inserted femoral venous sheaths.[22,23]

Our first choice is to perform balloon septostomy via the umbilical venous route. Therefore, we encourage our neonatology colleagues to place an umbilical venous line early on, with its tip well into the right atrium, before the ductus venosus constricts. At the time of septostomy, this line is exchanged over a wire with an appropriate sized sheath.

Initially Rashkind balloon septostomy catheters (USCI, Boston, MA) were used. Because the catheters were straight, sometimes making it difficult to advance the catheter into the left atrium, and because of the limited volume of fluid that these balloons would take, most cardiologists
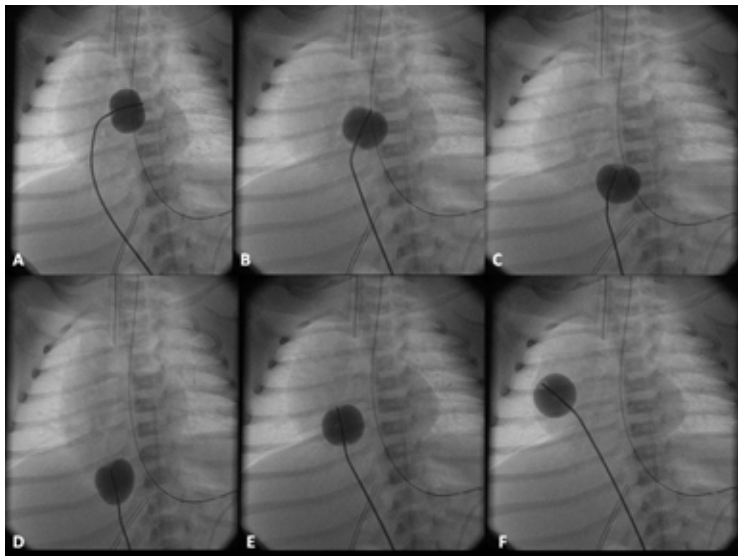
Figure 8. Selected cinfluroscopic frames of the Rashkind's balloon septostomy procedure. Note the position of the inflated balloon in the left atrium (A) and in right atrium and inferior vena cava in successive frames, as it is rapidly and forcefully withdrawn across the atrial septum (B,C, & D). After it reaches the inferior vena cava (D), it is rapidly advanced into the right atrium (E & F) in order not to inadvertently occlude the inferior vena cava in case of failure to deflate the balloon (which is quite rare).

have switched to Edwards septostomy catheters (American Edwards Baxter, McGaw Park, IL). These catheters have a gentle curve at the tip, facilitating easy access into the left atrium and larger volume of fluid that can be injected into these balloons. More recently, atrioseptostomy catheters (B/Braun, Bethlehem, PA) have become available. There are no studies comparing the relative effectiveness of the available catheters and, therefore, the selection of the type of catheter used is at the discretion of the operator.

The feasibility of performing balloon septostomy bedside, under echo guidance, has been demonstrated.[24,25] But, most cardiologists perform the procedure in the catheterization laboratory which is preferred by the author.

TGA with VSD patients usually present with heart failure and aggressive anti-congestive measures are indeed needed. Balloon atrial septostomy may help relieve pulmonary venous congestion and improve oxygenation. These patients will require Jatene procedure along with closure of the VSD.

TGA with VSD and PS patients may have varying types of presentation. If poor mixing is the reason for hypoxemia, balloon atrial septostomy is the treatment of choice. If the hypoxemia is secondary to markedly decreased pulmonary flow, a Blalock-Taussig type of shunt[26,27] may be needed. Sometimes both transcatheter balloon atrial septostomy and balloon pulmonary valvuloplasty[28-31] may be needed to improve hypoxemia. Most of these patients eventually require a Rastelli type of repair.[32]

Surgical Correction

Two types of surgical approaches, namely atrial (venous) and arterial switch are available for use. In the venous switch procedure, the systemic venous flow is directed towards the mitral valve and the pulmonary venous flow towards the tricuspid valve by constructing an intra-atrial baffle after the removal of the atrial septum. This is a physiological (hemodynamic) correction reversing the blood flow pathways at entry to counter the congenitally reversed great arteries. But the procedure leaves the morphological right ventricle to pump

against the high resistance systemic circuit. Originally described by Mustard in 1964,[33,34] the procedure was the most commonly used operation for TGA in the past. Similar venous re-directing procedures described by Senning[35] and Shumacker[36] have also been used in several centers. While post-operative complications such as arrhythmia and baffle obstruction were reduced significantly by better understanding of the conduction system and its blood supply coupled with the use of a pericardial baffle (instead of Dacron baffle), they still remained significant. Furthermore, leaving morphological right ventricle to pump into the aorta caused right ventricular failure in adolescents and adults. Jatene et al[10] described anatomical corrections for TGA in 1975; they switched the aorta and pulmonary artery with relocation of the coronary arteries to the neo-aortic root. Such procedures were initially performed for TGA with non-restrictive VSD where the left ventricular pressure was at systemic level. Subsequently the procedure was adapted to TGA with intact septum. However, arterial switch procedure must be performed in the early neonatal period prior to deconditioning of the left (pulmonary) ventricle. The arterial switch procedure has several advantages when compared with the venous switch procedure in that the arrhythmias are less frequent, and the morphological left ventricle rather than the right ventricle serves as a pump for the systemic circulation. Although there are no extensive long-term follow-up results available, the short-term and medium-term follow-up results are very encouraging and, at this time, the arterial switch procedure with or without LeCompte maneuver is considered the preferable operation for patients with TGA. Follow-up after surgery is mandatory to detect and manage residual defects.

Group III TGA patients with VSD and PS most often require Rastelli type of surgery[32] in which left ventricular blood flow is directed into the aorta with the VSD closing patch and a valved conduit (usually an aortic homograft) is inserted to connect the right ventricle to the pulmonary artery. This type of "corrective" surgery is not usually performed during the neonatal period and therefore, will not be discussed further. Palliation may be required during the neonatal period, as discussed in the preceding section.

SUMMARY AND CONCLUSIONS

Transposition of the great arteries is a congenital heart defect in which the aorta arises from the right ventricle, while the pulmonary artery comes off the left ventricle. It is the most common cyanotic CHD in the neonate. In this condition the systemic and pulmonary circulations are parallel instead of the normal circulation which is in series. This anomaly is classified into TGA with intact ventricular septum, VSD and VSD with PS. The intact ventricular septum patients present in the very early neonatal period while the other two may present with symptoms slightly later. Cyanosis is the major symptom in intact septum patients, while heart failure is the presenting symptom in patients with TGA and VSD. TGA with VSD and PS have a variable presentation. Murmurs are notably absent in intact septum babies while loud holosystolic or ejection systolic murmurs dominate in the other two groups. While the chest x-ray and ECG are helpful in the diagnosis, echocardiographic studies are confirmatory in the diagnosis and quantification of the associated defects. PGE_1 to open the ductus and/or balloon atrial septostomy to enlarge the PFO may sometimes be required for palliation. Corrective surgery by arterial switch (Jatene) procedure is necessary in TGA patients with intact septum and those with VSD whereas Rastelli procedure may be required for TGA patients with VSD and PS.

REFERENCES

1. Rao PS. Perinatal circulatory physiology: It's influence on clinical manifestations of neonatal heart disease – Part I. Neonatology Today 2008; 3(2):6-12.
2. Rao PS. Perinatal circulatory physiology: It's influence on clinical manifestations of neonatal heart disease – Part II. Neonatology Today 2008; 3(3):1-10.
3. Rao PS. An approach to the diagnosis of cyanotic neonate for the primary care provider. Neonatology Today 2007; 2 (6):1-7.
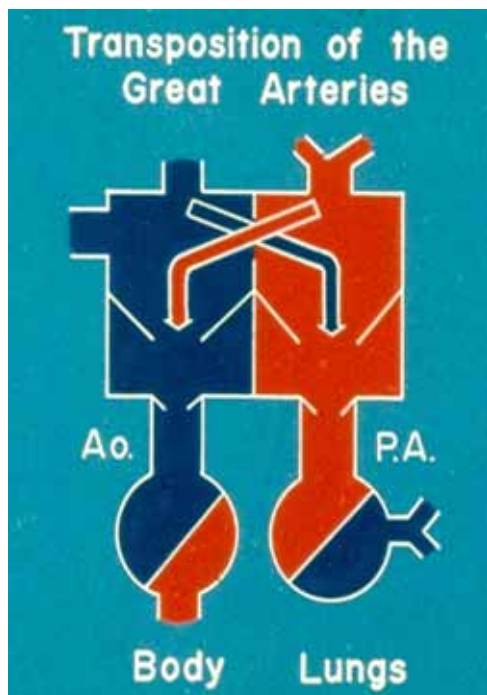
Figure 9. Diagrammatic representation of left-to-right (pink arrow) and right-to-left (blue arrow) shunt across the patent foramen ovale which has been enlarged by balloon atrial septostomy. Ao., aorta; P.A., pulmonary artery.

4. Rao PS. Principles of management of the neonate with congenital heart disease Neonatology Today 2007; 2(8): 1-10.
5. Rao PS. Neonatal cardiac emergencies: Management strategies, Neonatology Today 2008; 3(12):1-5.
6. Fyler DC (ed): Nadas' Pediatric Cardiology, Hanley & Belfus, Inc., Philadelphia, PA, 1992.
7. Rao PS. Diagnosis and management of cyanotic congenital heart disease: Part I. Indian J Pediat 2009; 76:57-70.
8. Rashkind WJ, Miller WW. Creation of an atrial septal defect without thoracotomy. J Am Med Assoc 1966; 196:991-2.
9. Rao PS. Role of interventional cardiology in neonates: Part I. Non-surgical atrial septostomy. Neonatology Today 2007; 2(9):9-14.
10. Jatene AD, Fontes VF, Paulista PP, et al. Anatomic correction of transposition of the great vessels. J Thorac Cardiovasc Surg 1976; 72:364-70.
11. Rao PS. Static balloon dilation of atrial septum (Editorial). Am Heart J 1993; 125: 1826-7.
12. Park SC, Neches WH, Zuberbuhler JR, et al. Clinical use of blade septostomy. Circulation 1978; 58: 600-6.
13. Shrivatsava S, Radhakrishnan S, Dev V, et al. Balloon dilatation of atrial septum in complete transposition of great arteries - a new technique. Indian Heart J 1987; 39:298-300.
14. Rao PS. Static balloon dilation of restrictive atrial septal defects (Editorial). J Saudi Heart Assoc 1992; 4: 55-8.
15. Gewillig D, Boshoff L, Mertens L. Creation with a stent of an unrestrictive lasting atrial communication, Cardiol Young 2002; 12:404–7.
16. Eicken H, Gildein, C, Schreiber C, et al. Stenting of a restrictive foramen ovale in a patient with hypoplastic left heart syndrome. Internat J Cardiol, 2006; 113: 254-6A.
17. Atz AM, Feinstein JA, Jonas RA, et al. Preoperative management of pulmonary venous hypertension in hypoplastic left heart syndrome with restrictive atrial septal defect. Am J Cardiol 1999; 83: 1224–8.
18. Justino H, Benson LN, Nykanen DG. Transcatheter creation of an atrial septal defect using radiofrequency perforation. Catheter Cardiovasc Intervent. 2001; 54:83-7.
19. Sakata Y, Feldman T. Transcatheter creation of atrial septal perforation using a radiofrequency transseptal system: novel approach as an alternative to transseptal needle puncture. Catheter Cardiovasc Intervent. 2005; 64:327-32.
20. Hill SL, Mizelle KM, Vellucci SM, et al. Radiofrequency perforation and cutting balloon septoplasty of intact atrial septum in a newborn with hypoplastic left heart syndrome using transesophageal ICE probe guidance, Catheter Cardiovasc Intervent 2005; 64:214–7.
21. Abinader E, Zeltzer M, Riss E. Transumbilical atrial septostomy in the newborn. Am J Dis Child 1970; 119:354-5.
22. Hurwitz RA, Girod DA. Percutaneous atrial septostomy in infants with transposition of the great arteries. Am Heart J 1976; 91:618-22.
23. Sunderland CO, Nichols GM, Henken DP, et al. Percutaneous cardiac catheterization and atrial balloon septostomy in pediatrics. J Pediat 1976; 89:584-7.
24. Baker EJ, Allan LD, Tynan M, et al. Balloon atrial septostomy in the neonatal intensive care unit. Br Heart J 1984; 51:377-8.
25. Bullaboy CA, Jennings RB, Jr, Johnson DH. Bedside balloon atrial septostomy using echocardiographic monitoring. Am J Cardiol 1984; 53:971-2.
26. Blalock A, Taussig HB. The surgical treatment of malformations of the heart in which there is pulmonary stenosis or atresia. JAMA 1945; 128:189-202.
27. de Leval MR, McKay R, Jones M, et al. Modified Blalock-Taussig shunt. Use of subclavian artery orifice as flow regulator in prosthetic systemic-pulmonary artery shunts. J Thorac Cardiovasc Surg 1981; 81:112-9.
28. Rao PS and Brais M. Balloon pulmonary valvuloplasty for congenital cyanotic heart defects. Am Heart J 1988; 115:1105-10.
29. Rao PS, Wilson AD, Thapar MK, Brais M. Balloon pulmonary valvuloplasty in the management of cyanotic congenital heart defects. Cathet Cardiovasc Diagn 1992; 25:16-24.
30. Rao PS. Transcatheter management of cyanotic congenital heart defects: a review. Clin Cardiol 1992; 15:483-96.
31. Rao PS. Pulmonary valve in cyanotic heart defects with pulmonary oligemia. In: Percutaneous Interventions in Congenital Heart Disease, Sievert H, Qureshi SA, Wilson N, Hijazi Z (Eds), Informa Health Care, Oxford, UK, 2007, pp 197-200.
32. Rastelli GC, McGoon DC, Wallace RB. Anatomic correction of transposition of the great arteries with ventricular septal defect and subpulmonary stenosis. J Thorac Cardiovasc Surg 1969 ; 58:545-52.
33. Mustard WT. Successful two-stage correction of transposition of the great vessels. Surgery. 1964; 55:469-72.
34. Mustard WT, Keith JD, Trusler GA, et al. The surgical management of transposition of the great vessels. J Thorac Cardiovasc Surg 1964 ; 48:953-8.
35. Senning A. Surgical correction of transposition of the great vessels. Surgery 1959; 45:966-80.
36. Alvarado A. Modified Shumacker operation for correction of transposition of the great arteries. J Thorac Cardiovasc Surg. 1977; 74:614-7.

CCT

P. Syamasundar Rao, MD
Professor and Director,
Division of Pediatric Cardiology
UT-Houston Medical School
6410 Fannin, UTPB Ste. #425
Houston, TX 77030 USA

Phone: 713-500-5738
Fax: 713-500-5751

P.Syamasundar.Rao@uth.tmc.edu

Review of the Congenital Heart Disease Program at SCAI, 2010

By Daniel Levi, MD and Frank Ing, MD

With proximity to the bay, beach, Gas Lamp District and Petco Park, the modern style of San Diego's Hilton Bayfront Hotel provided an excellent setting for the *33rd Annual SCAI (The Society for Cardiovascular Angiography and Interventions)* held in early May 2010. This year, the Congenital Heart Disease Program was expanded to two and a half days. A host of new sessions included: "Brain Scratchers," "When Devices Fail," "Old Arts and New Arts" and "Mythbusters." This collection of sessions was chosen to encourage presentation of creative and atypical cases and topics not always considered at pediatric interventional catheterization meetings. Topics were designed to stimulate discussion from the audience.

One of the most successful sessions was the new "Brain Scratchers" session that allowed for presentation of challenging cases that raised puzzling questions for the audience. These cases were not necessarily "nightmare cases." Each speaker challenged the audience to solve hemodynamic, angiographic or interventional mysteries or to provide solutions for less than routine cases in the congenital catheterization laboratory. The "Brain Scratchers" session was highlighted by cases from: Dr. Larry Latson (the Cleveland Clinic, who showcased a patient with differential flow from a Glenn shunt causing severe cyanosis and a creative treatment plan that ultimately normalized his oxygen saturation); Dr. Lee Benson (Toronto Sick Kids, case of a pulmonary angioplasty gone bad); Dr. Henri Justino (Texas Children's Hospital, a difficult pulmonary venous obstruction) and from Dr. Dennis Kim (Sibley Heart Center in Atlanta, case of "mystery cause of persistent collaterals"). Lee Benson even asked the fellows to move to the front row and gave them a taste of "the Toronto treatment." This session provided an excellent forum other than the "I Blew It" session to allow for audience participation. Nearly everyone in the room got involved with creative suggestions for these interesting case presentations. The grand finale of the session was given by Dr. Evan Zahn (Miami Children's). In his own unique style, Evan presented a brain scratching case of a large stent loose in the right ventricle.

New "Mythbusters" session included a talk by Dr. Henri Justino on "The Congenital Absent Pulmonary Artery." Henri demonstrated the benefits of looking very carefully for pulmonary arteries in cases in which the pulmonary arteries are "claimed" to be "congenitally absent." In many of these cases, "absent" arteries can be found and grown into substantial pulmonary arteries if discovered and unifocalized early. Dr. Seong-Ho Kim from Korea showed cases of PDA closure in the setting of pulmonary hypertension and Dr. Evan Zahn showed cases in which ASDs with absent rims can be safely closed. Dr. Lourdes Prieto (Cleveland Clinic) discussed strategies for recanalizing occluded pulmonary veins. Finally, Dr. Frank Ing (Texas Children's Hospital) dispelled the myth that the subcostal approach is the only safe way to drain a pericardial effusion. Dr. Ing presented his experience with other safe approaches for pericardiocentesis.

Dr. Robert Vincent (Emory University) had the hardest job of the *2010 SCAI*… he was asked to face Dr. Welton Gersony (Columbia University) in the debate over "Should we close the silent ductus?" Although Dr. Vincent put up a valiant effort on the side of favoring closure, Dr. Gersony made some very convincing arguments against closure of very small PDAs. While the audience overwhelmingly favored not closing the very small PDA in the cath lab, this simple topic sparked an excellent analysis of the real risks of small PDAs in the general population in the modern era.

The first annual Mullins Lecture was given by Dr. John W. Moore (Rady Children's Hospital of San Diego). Dr. Moore is one of the many Mullins'

Left to right: Drs. Charles Mullins, Frank Ing, and Welton Gersony.

Left to right: Drs. Charles Mullins, Mario Carminati, Donald Hagler, and John P. Cheatham.

Left to right: Drs. Jeff Darst, Lourdes Prieto, Thomas Fagan and Daniel Levi.

trained pediatric interventionalists now leading the field of pediatric interventional cardiology. Dr. Moore talked about the past and future of stent technology. Dr. Mullins' contribution and pioneering work in stent applications for congenital heart disease was featured prominently.

The "When Devices Fail" session provided a forum to discuss issues involving occlusion devices, stents and transcatheter valves. Dr. Thomas Fagan (Children's Hospital of Denver) discussed the many ways in which stents can fail and fracture and highlighted the testing done on stents to avoid failure. Dr. Ziyad Hijazi (Rush University) and Dr. Mario Carminati from Italy discussed old and new failure issues with the Sappien and Melody valves respectively. Dr. Zahid Amin (Rush

Left to right: Drs. Daniel Levi, John W. Moore, John P. Cheatham and Charles Mullins discussing a presentation.

Left to right: Dr. Lourdes Prieto, Dr. & Mrs. Kak-Chen Chan, and Dr. & Mrs. Larry Latson.

Left to right: Dr. Ahmed Alomrani, Dr. & Mrs. Frank Ing and Dr. Seong-Ho Kim at the SCAI Gala.

Dr. Ahmed Alomrani, winner of the "I Really Blew It" award

transcatheter devices. His talk highlighted the use of thin film nitinol and other smart materials in pediatric devices.

The "I Blew It" sessions (11th year) once again both shocked, entertained and educated all of us on the various ways in which interventional cases can go awry, and novel and creative ways to manage these complications. The "I Really Blew It!" award was voted to Dr. Ahmed Alomrani from Riyadh who presented a case of a LPA stent case "gone wild."

Finally, eight excellent oral abstracts were presented over the course of the meeting. The entire *2010 SCAI* was well-attended and very successful on all fronts. The 34th session of the *SCAI* will be held in Baltimore, MD from May 4-7, 2011 (www.scai.org/SCAI2011). The congenital program is already being developed, and promises to maintain the quality established by the 2010 program. Hope to see you there.

CCT

University) and Dr. Lee Benson provided demonstrations and reminders of the ways in which ASD and PDA devices can fail us. While each talk stressed mechanisms of failure, each speaker also outlined strategies for avoiding these failures.

In the final session entitled "Lost and New Arts," Dr. Charles Mullins gave an excellent talk on the lost art of gathering precise hemodynamic data and catheter manipulation. His lecture was followed by a talk on newer hybrid procedures "Beyond HLHS" in which Dr. John Cheatham (Columbus Children's Hospital) showed the range of awesome new procedures that he has been able to perform with his surgeons in a hybrid fashion. In the final lecture, Dr. Daniel Levi (Mattel Children's Hospital at UCLA) discussed use of novel materials for the design of a new breed of

Daniel S. Levi, MD
Mattel Children's Hospital at UCLA
Pediatric Cardiology, B2-427 MDCC
C310 200 5003
10833 Le Conte Ave.
Los Angeles, CA 90095-1743 USA
Phone: 310.206.3478
Pager: 1.800.233.7231 page ID#20347
dlevi@ucla.edu

Frank Ing, MD
Texas Children's Hospital
6621 Fannin St.
MC 19345-C
Houston TX 77030 USA
fxing@texaschildrens.org

Abstracts from "Evolving Concepts in the Management of Complex Congenital Heart Disease II" - Part V

Abstract Title: The Infant Ross / Konno: Early and Late Results
Presenter: Vaughn A. Starnes, MD; Head, Division of Cardiothoracic Surgery; Children's Hospital Los Angeles, Los Angeles, CA USA

Objective

1. Describe one benefit of the Ross Procedure.
2. Identify the controversy surrounding the Ross Procedure.

Abstract

Infants with critical aortic stenosis with or without left ventricular outflow tract obstruction (LVOTO) present a unique surgical challenge. The Ross Procedure was pioneered by Donald Ross, MD in 1967 in the adult population, and rapidly became an option for the treatment of aortic valve disease in children. Increasingly, the Ross and/or Ross/Konno is being utilized in the infant population, with good results. The benefits of the Ross Procedure include:
1. Potential for growth of the autograft.
2. Avoidance of the need for anticoagulation.
3. Appropriateness of valvar size for infants.

There are also disadvantages with the Ross Procedure in infants, and some would argue that the operation turns single valve disease into double valve disease.[2] There is the issue of interventions in the right ventricular outflow tract due to pulmonary valve replacement, and recently in the literature there have been multiple reports of dilation of the neoaortic root, leading to regurgitation.[1]

In our experience at the Children's Hospital Los Angeles, 174 children have received the Ross Operation (1993-2009). Of these 174 children, 33 have been infants with critical aortic stenosis less than 12 months of age, and that is the population which will be discussed at this meeting.

Review of 33 patients <12 months of age at CHLA, who underwent the Ross or Ross/Konno can be categorized as follows:
1. Urgent.
2. Patients who were hospitalized in the Intensive Care Unit, requiring supportive care.
3. Elective.
4. Patients followed on an Outpatient basis.

Of the 33 infants in this series the median age at operation was 3.8 months (2 days to 317 days of age at surgery). There were 11 females and 22 males. Hospital mortality is defined as death prior to hospital discharge post Ross procedure, and was 18% (6/33). Overall mortality is 18% (6/33) with 0 deaths occurring after discharge from the hospital. Of the 33 infants, 17 patients underwent a Ross/Konno, and 16 underwent Ross Procedure. Forty-two percent of the patients had at least one intervention prior to the Ross Procedure (14/33). Eight patients (24%) had mitral valve disease, and 7 patients underwent concomitant mitral valvuloplasty (5 Ross Konno and 2 Ross), with one undergoing mitral valve replacement as salvage 3 weeks post Ross Konno. Two patients had fibroelastosis (1 Ross Konno and 1 Ross). Of the 27 surviving patients, 44% have had their conduit replaced (12/27), and one was operated at 4 years post Ross for subaortic membrane.

The Ross Procedure provides an acceptable option in dealing with the patient with critical aortic stenosis and should be considered as a treatment option.

References

1. Alexander Kadner, Olivier Raisky, Alexandra Degandt, Daniel Tamisier, Damien Bonnet, Daniel Sidi, Pascal R. Vouhé The Ross Procedure in Infants and Young Children Ann. Thorac. Surg., March 2008; 85: 803-808.
2. Richard G Ohye, Carlen A Gomez, Bonita J Ohye, Caren S Goldberg, Edward L Bove The Ross/Konno procedure in neonates and infants: intermediate-term survival and autograft function Ann Thorac Surg., September 2001; 72: 823-830.
3. Sara K. Pasquali, Bradley S. Marino, Jonathan R. Kaltman, Andrew J. Schissler, Gil Wernovsky, Meryl S. Cohen, Thomas L. Spray, Ronn E. Tanel Rhythm and Conduction Disturbances at Midterm Follow-up After the Ross Procedure in Infants, Children, and Young Adults Ann Thorac Surg., June 2008, 85: 2072-2078.

CCT

Abstract Title: Management of Children with Aortic or Mitral Regurgitation: Role of Medical Therapy and Timing of Surgery
Presenter: Lloyd Y. Tani, MD
Chief, DIvision of Pediatric Cardiology, University of Utah School of Medicine, Primary Children's Medical Centers, Salt Lake City, UT USA

Objective

1. To review the role of medical management in chronic MR.
2. To review indications for surgical intervention in chronic MR.
3. To review the role of medical management in chronic AR.
4. To review the indications for surgical intervention in chronic AR.

Abstract

I. Guidelines
    A. 2008 Focused Update of ACC/AHA 2006 Guidelines for the Management of Patients with Valvular Heart Disease .
    B. Guidelines are just guidelines.
    C. Lack of randomized controlled trials.
    D. Classes of recommendations.
II. Principles influencing recommendations.
    A. Limited data in children.
    B. Natural history of severe MR and AR in children.
    C. Extrapolate from adult studies.
    D. Goals of treatment (medical or surgical).
III. Mitral Regurgitation.
    A. Approach to Patient.
    B. Role of Echocardiography.
        1. Etiology.
        2. Establish Severity.
        3. LV size and function.
    C. Role of BNP.
    D. Medical Management.
        1. Is there a role for medical management?
    E. Timing of Surgery.
        1. Factors influencing Outcome.
        2. Indications.
            a) Symptoms.
            b) Marked LV dilation.
            c) LV dysfunction.
            d) Atrial fibrillation or pulmonary hypertension.
            e) What about the asymptomatic patient without marked LVE or LV dysfunction.
                (1) Importance of likelihood of repair vs. replacement.
            f) What do we know about children?
            g) Aortic Regurgitation.
IV. Aortic Regurgitation.
    A. Approach to Patient.
    B. Role of Echocardiography.
        1. Etiology.
        2. Establish Severity.
        3. LV size and function.
    C. Medical Management.
        1. Role for afterload reduction?.
        2. Conflicting Evidence.
    D. Timing of Surgery.
        1. Factors influencing outcome.
        2. Indications.
            a) Symptoms.
            b) Marked LV dilation.
            c) LV dysfunction.
            d) What do we know about children?

References

1. Bonow, R. O., B. A. Carabello, et al. (2008). "2008 Focused update incorporated into the ACC/AHA 2006 guidelines for the management of patients with valvular heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines (Writing Committee to Revise the 1998 Guidelines for the Management of Patients With Valvular Heart Disease): endorsed by the Society of Cardiovascular Anesthesiologists, Society for Cardiovascular Angiography and Interventions, and Society of Thoracic Surgeons." Circulation 118(15): e523-661.
2. Carabello, B. A. (2008). "The current therapy for mitral regurgitation." J Am Coll Cardiol 52(5): 319-26.
3. Enriquez-Sarano, M., C. W. Akins, et al. (2009). "Mitral regurgitation." Lancet 373(9672): 1382-94.
4. Stewart, W. J. (2006). "Optimal timing of surgery in aortic regurgitation." Heart Fail Clin 2(4): 461-71.
5. Bermudez, E. A. and W. H. Gaasch (2006). "Regurgitant lesions of the aortic and mitral valves: considerations in determining the ideal timing of surgical intervention." Heart Fail Clin 2(4): 473-82.

Tani Mgmt MRAR Abstract Management of Children with Aortic or Mitral Regurgitation: Role of Medical Therapy and Timing of Surgery

CCT

Abstract Title: Role of Echocardiography in the Evaluation of Mitral and Aortic Regurgitation
Presenter: Lloyd Y. Tani, MD
Chief, DIvision of Pediatric Cardiology, University of Utah School of Medicine, Primary Children's Medical Centers, Salt Lake City, UT USA

Objective

1. Review the role of echocardiography in the evaluation of mitral regurgitation.
2. Review the echocardiographic assessment of MR severity.
3. Review the role of echocardiography in the evaluation of aortic regurgitation.
4. Review the echocardiographic assessment of AR severity.

Abstract

I. Assessment of Mitral and Aortic Regurgitation.

A. Importance of History and Physical.
B. Central Role of Echocardiography.
C. Importance of using results in the context of the patient.
II. Echocardiographic Assessment of Mitral Regurgitation.
  A. Diagnoses Associated with MR in children.
  B. Why echo assessment of MR important.
    1. Documentation.
    2. Assessment of severity.
    3. Progression may occur.
    4. Central to management and determining timing of intervention.
  C. Parameters.
    1. Color Doppler size.
    2. Pulmonary Venous Doppler.
    3. Regurgitant volume, fraction, and orifice area.
      a) Doppler and 2D methods.
      b) Proximal isovelocity surface area (PISA).
    4. Size of vena contracta.
    5. Severity index.
    6. Continuous wave Doppler.
    7. LV size and function.
  D. Assessment of valve morphology.
    1. Repair vs. Replacement.
III. Echocardiographic Assessment of Aortic Regurgitation.
  A. Diagnoses associated with AR in children.
  B. Why echo assessment of AR important.
    1. Documentation.
    2. Assessment of severity.
    3. Progression may occur.
    4. Central to management and determining timing of intervention.
  C. Parameters.
    1. Color jet size.
      a) Jet width.
      b) Short axis jet area.
    2. Regurgitant volume, fraction, and orifice area.
    3. Continuous wave Doppler.
    4. Flow in the downstream aorta.
    5. LV size and function.
  D. Assessment of valve morphology.
    1. Repair vs. Replacement.

References

1. Zoghbi, W. A., M. Enriquez-Sarano, et al. (2003). "Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography." J Am Soc Echocardiogr 16(7): 777-802.
2. Thomas, L., E. Foster, et al. (1999). "The Mitral Regurgitation Index: an echocardiographic guide to severity." J Am Coll Cardiol 33(7): 2016-22.
3. Tani, L. Y., L. L. Minich, et al. (1997). "Doppler evaluation of aortic regurgitation in children." Am J Cardiol 80(7): 927-31.
4. Barrea, C., S. Levasseur, et al. (2005). "Three-dimensional echocardiography improves the understanding of left atrioventricular valve morphology and function in atrioventricular septal defects undergoing patch augmentation." J Thorac Cardiovasc Surg 129(4): 746-53.
5. Lim, D. S., J. M. Dent, et al. (2007). "Transesophageal echocardiographic guidance for surgical repair of aortic insufficiency in congenital heart disease." J Am Soc Echocardiogr 20(9): 1080-5.
6. Enriquez-Sarano, M., J. F. Avierinos, et al. (2005). "Quantitative determinants of the outcome of asymptomatic mitral regurgitation." N Engl J Med 352(9): 875-83.
7. Tribouilloy, C. M., J. F. Avierinos, et al. (2004). "Impact of echocardiography on indications for surgery in chronic mitral and aortic regurgitation." Clin Cardiol 27(8): 442-8.

CCT

Abstract Title: It's Hard to Top a Well-Done Norwood
Presenter: James S. Tweddell, MD
The S. Bert Litwin Chair; Cardiothoracic Surgery; University of California, San Diego School of Medicine, San Diego, CA USA

Objective:

- Review Norwood operative techniques that minimize the risk or recurrent or residual lesions.
- Review principles of goal directed postoperative management.
- Review interstage monitoring and the impact on interstage mortality.

Abstract:

The goals of the Norwood procedure are well understood: provide unobstructed systemic outflow from the single ventricle including coronary blood flow, limitation of excessive pulmonary blood flow using an expanded PTFE graft and prevention of pulmonary venous hypertension by creation of an unrestrictive atrial septal defect.[1] The Norwood procedure requires cardiopulmonary bypass and altered perfusion and the postoperative course is long and includes the need for inotropic support and prolonged mechanical ventilation. A well-done Norwood takes advantage of techniques that limit recurrent or residual lesions that add to the hemodynamic burden, increase vulnerability and result in excess morbidity and mortality. Optimal arch reconstruction includes an interdigitating anastomosis that allows for growth and essentially eliminates the risk of recurrent obstruction.[2,3] Regardless of the choice of shunt, optimal shunt construction must include initial selection of the proper caliber shunt and techniques that minimize the risk of shunt stenosis. Postoperative care should include measures of systemic oxygen delivery and interstage monitoring, necessary to provide early detection of recurrent/residual lesions and intercurrent illness that can result in mortality.[4,5] The advantage of a well-done Norwood procedure is the early and complete elimination of excessive hemodynamic burden that will permit optimal long-standing single ventricle function.

References:

1. Tweddell JS. The Norwood Procedure with an Innominate Artery-to-Pulmonary Artery Shunt. Operative Techniques in Thoracic and Cardiovascular Surgery. 2005 7:123-140.
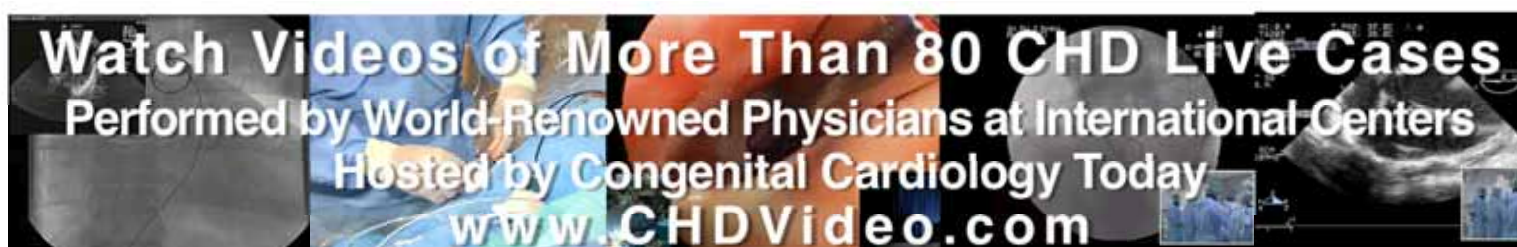
2. Lamers L, Frommelt PC, Mussatto KA, Mitchell ME, Tweddell JS. Decreased Incidence of Recurrent Coarctation of the Aorta Following the Norwood Procedure. Circulation. 2007;116:II_515.
3. Burkhart HM, Ashburn DA, Konstantinov IE, De Oliveira NC, Benson L, Williams WG, Van Arsdell GS. Interdigitating arch reconstruction eliminates recurrent coarctation after the Norwood procedure. J Thorac Cardiovasc Surg. 2005;130:61-5.
4. Tweddell JS, Hoffman GM, Ghanayem NS, Frommelt MA, Mussatto KA, Berger S. Hypoplastic Left Heart Syndrome. In, Moss and Adams' Heart Disease in Infants, Children, and Adolescents : Including the Fetus and Young Adult. 7th Ed. Allen HD, Driscoll DJ, Shaddy RE, Feltes TF (eds). Lippincott, William and Wilkins Publishers, Philadelphia, PA, October 2007.
5. Ghanayem NS, Cava JR, Jaquiss RD, Tweddell JS. Home monitoring of infants after stage one palliation for hypoplastic left heart syndrome. Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu. 2004;7:32-8.

CCT

Abstract Title: Treatment of AET and JET – Native and Post-Op
Presenter: George F. Van Hare, MD; Director, Division of Pediatric Cardiology; Washington University / St. Louis Children's Hospital; St. Louis, MO USA

Objective:

To be conversant with the medical treatment and the ablation techniques available for the treatment of atrial ectopic tachycardia and junctional ectopic tachycardia

Abstract:

Atrial ectopic tachycardia and junctional ectopic tachycardia are both seen in otherwise normal children, but are also seen in the immediate post-operative period following cardiac surgery for the repair of congenital cardiac defects.

In the situation of "native" AET and JET, the etiology of these conditions is unclear. AET may be the result of inapparent viral myocarditis. JET may have a similar etiology, but the familial occurrence suggests a more interesting etiology. Dubin et al. identified JET as a manifestation of antibody-mediated AV block in infants of mothers with lupus. Presumably, the JET is due to conduction system injury which is not severe enough to cause complete block. Both AET and JET respond to various antiarrhythmic medications. Spontaneous resolution is reported in both, and so ablation may not be needed. Catheter ablation is highly effective for both. With JET, of course, there is the risk of AV block, but this risk is remarkably small considering the fact that JET presumably arises from the AV node or His bundle. Cryoablation is a good choice in this substrate.

Post-operative AET is reported but is not common. It must of course be differentiated from atrial flutter. It is managed pharmacologically, with the expectation that it will resolve. Post-operative JET is more common, but there is a wide variation in the incidence by institution. It presumably is a result of perinodal injury and is highly associated with septal defect closure. Prevention is a tantalizing possibility – pretreatment with magnesium has been reported to be successful. The arrhythmia can be expected to resolve in the time frame of 1-2 days, but it may lead to hemodynamic instability. Non-pharmcological approaches may be effective, especially atrial overdrive pacing to restore AV synchrony, and cooling. A number of agents have been used to treat post-operative JET, including digoxin and procainamide. Most centers primarily employ intravenous amiodarone. There is essentially no role for catheter ablation in this situation.

The recent widespread adoption of the anesthetic agent dexmedetomidine (Precedex) for sedation following cardiac surgery may well be changing the landscape of JET. This agent has clear vagomimetic effects. Chrysostomou et al have reported its successful use to treat both JET and AET post-operatively.

References:

1. Walsh EP, Saul JP, Sholler GF, Triedman JK, Jonas RA, Mayer JE, Wessel DL. Evaluation of a staged treatment protocol for rapid automatic junctional tachycardia after operation for congenital heart disease. J Am Coll Cardiol. 1997;29(5):1046-1053.
2. Collins KK, Van Hare GF, Kertesz NJ, Law IH, Bar-Cohen Y, Dubin AM, Etheridge SP, Berul CI, Avari JN, Tuzcu V, Sreeram N, Schaffer MS, Fournier A, Sanatani S, Snyder CS, Smith RT, Jr., Arabia L, Hamilton R, Chun T, Liberman L, Kakavand B, Paul T, Tanel RE. Pediatric nonpost-operative junctional ectopic tachycardia medical management and interventional therapies. J Am Coll Cardiol. 2009;53 (8):690-697.
3. Dorman BH, Sade RM, Burnette JS, Wiles HB, Pinosky ML, Reeves ST, Bond BR, Spinale FG. Magnesium supplementation in the prevention of arrhythmias in pediatric patients undergoing surgery for congenital heart defects. Am Heart J. 2000;139(3):522-528.
4. Hammer GB, Drover DR, Cao H, Jackson E, Williams GD, Ramamoorthy C, Van Hare GF, Niksch A, Dubin AM. The effects of dexmedetomidine on cardiac electrophysiology in children. Anesth Analg. 2008;106 (1):79-83, table of contents.
5. Chrysostomou C, Beerman L, Shiderly D, Berry D, Morell VO, Munoz R. Dexmedetomidine: a novel drug for the treatment of atrial and junctional tachyarrhythmias during the perioperative period for congenital cardiac surgery: a preliminary study. Anesth Analg. 2008;107(5):1514-1522.

CCT

Medical News, Products and Information

BioSTAR™ Device Achieves 90% Closure Rate for Atrial Septal Defect in Children - Biodegradable Implant Avoids Risks Associated with Metal Devices

A novel study by Canadian physicians reported that the BioSTAR™ biodegradable implant achieved comparable closure rates to the Amplatzer Septal Occluder™ (ASO) in children with atrial septal defect (ASD). The BioSTAR device displayed successful outcomes, while avoiding issues associated with implants containing substantial amounts of metal. Results of the study, the first to compare the BioSTAR device with the ASO in children, are now available online and in the July print issue of Catheterization and Cardiovascular Interventions, a journal published by Wiley-Blackwell on behalf of The Society for Cardiovascular Angiography and Interventions.

ASD is a congenital heart defect where a hole is present in the wall between the two upper heart chambers. While smaller ASDs may close on their own in early life, larger defects may require surgery to close the opening. If left untreated, ASD may increase the risk of developing atrial fibrillation, heart failure, or stroke later in life. The incidence of atrial septal defects ranges from one-twentieth to one-tenth of all congenital heart lesions.

During the study period of November 1, 2007 through November 30, 2008 there were 54 children who underwent ASD closure with the ASO implant. The ASO group subjects' median data were: age of 7.4, weight of 23.3 kg, defect size of 10 mm, and balloon stretched size of 12.7 mm. The BioSTAR implant was used in 10 patients where a small to moderate defect was anticipated by non-invasive studies. In this group, patients' media data were: age of 11, weight of 39.6 kg, defect size of 10 mm, and balloon stretched size of 11.5 mm.

Study results indicated the acute and 6-month follow-up closure rate for the BioSTAR were both 90%, compared with 100% for both time periods with the ASO implants. "Our study provides evidence that the BioSTAR implant achieves comparable closure rates to the ASO device in small-to moderate-ASD," said lead study author Lee Benson, MD, FSCAI. "Minimal foreign material remains after 6 months with the biodegradable implant, reducing the risks associated with devices containing significant amounts of metal." The study team noted that decreased long-term thrombogenicity, preserved transseptal access, decreased inflammatory response, and reduced potential of arrhythmogenicity and erosion were benefits of using biodegradable implants.

The team reported no serious complications in either group. However, statistically significant differences in the media procedure time (BioSTAR-52 minutes; ASO device-39.5 minutes) and fluoroscopy times (BioSTAR-6.7 minutes; ASO device-6.1 minutes) were observed. "Longer procedure and fluoroscopy times are a drawback of the BioStar implant, but should improve with familiarity with the device and deployment system," Dr. Benson concluded.

Article: "A Biodegradable Device (BioSTAR™) for Atrial Septal Defect Closure in Children." Gareth Morgan, Kyong-Jin Lee, Rajiv Chaturvedi, and Lee Benson. Catheterization and Cardiovascular Interventions; Published Online: XX 2010 (DOI: 10.1002/ccd.22517); Print Issue: July 2010.

This study was published in *Catheterization and Cardiovascular Intervention*.

Catheterization and Cardiovascular Interventions is the official journal of Society for Cardiovascular Angiography and Interventions (SCAI).

Wiley-Blackwell is the international scientific, technical, medical, and scholarly publishing business of John Wiley & Sons, Inc., with strengths in every major academic and professional field and partnerships with many of the world's leading societies. Wiley-Blackwell publishes nearly 1,500 peer-reviewed journals and 1,500+ new books annually in print and online, as well as databases, major reference works and laboratory protocols. For more information, please visit www.wileyblackwell.com or www.interscience.wiley.com.

The Society for Cardiovascular Angiography and Interventions (SCAI) is the primary professional association for invasive and interventional cardiologists, representing over 4,300 physicians in 60 countries. The Society's mission is to promote excellence in invasive and interventional cardiovascular medicine through physician education and representation, its monthly journal Catheterization and Cardiovascular Interventions, and the advancement of quality standards to enhance patient care. For more information, please visit www.scai.org.

In Infant Heart Surgery, Newer Technique Yields Better Survival in First Year of Life

Pediatric researchers report that a recently introduced surgical procedure offers infants with severely underdeveloped hearts a better chance at surviving during their first year of life, in comparison to the standard surgery.

Heart surgeons from 15 centers in the federally sponsored Pediatric Heart Network studied the outcomes in 549 newborns who received a complex series of surgeries for Hypoplastic Left Heart Syndrome (HLHS).

"This landmark study is the largest clinical trial ever performed in congenital heart surgery, and the first randomized trial comparing two surgical procedures for congenital heart defects," said senior author J. William Gaynor, MD, a pediatric cardiothoracic surgeon at The Children's Hospital of Philadelphia, and co-principal investigator of the study. As one of the nation's leading programs in pediatric cardiology, the Cardiac Center at Children's Hospital enrolled 101 subjects in this study.

The study results appeared in the May 27th issue of the *New England Journal of Medicine*.

The lead author and principal investigator of this study, called the Single Ventricle Reconstruction (SVR) Trial, was Richard G. Ohye, MD, Head of the Pediatric Cardiovascular Surgery Division of the University of Michigan.

Occurring about once in every 100 live births, congenital heart disease is the most common birth defect. It includes a broad variety of structural abnormalities, but HLHS is among the most

severe forms. In HLHS, affecting 1 in 5,000 live births, the left ventricle, one of the heart's two pumping chambers, is small and unable to function. Without treatment, HLHS is fatal in the first few days of life.

Starting in the 1980s, surgeons developed surgical procedures for HLHS that have allowed increasingly more children born with a single functioning ventricle to survive. The intervention involves three planned surgeries, beginning in the newborn period and extending to 18 to 36 months of age.

The SVR trial reported in the current study compares two techniques used in the initial, riskiest stage of surgery, called the Norwood procedure. As part of the Norwood procedure, surgeons implant a shunt to reroute blood from the malformed heart to the pulmonary artery, which supplies the lungs.

The traditional surgical approach is to use a modified Blalock-Taussig (MBT) shunt, which carries blood from an artery branching off the aorta to the pulmonary artery. The newer technique, sometimes known as the Sano procedure or the right ventricle-pulmonary artery (RVPA) shunt, links the right ventricle to the pulmonary artery. If either technique fails, the only alternative is a heart transplant.

In the current trial, researchers randomized infants who required the Norwood procedure into two groups, 275 for the MBT shunt and 274 for the RVPA shunt. Twelve months after the surgery, 74% of infants with the RVPA shunt survived and didn't require a heart transplant, compared to 64% of infants with the MBT shunt. The RVPA group did, however, undergo a higher rate of complications requiring unintended interventions, such as needing stents or balloons to keep the shunt open. After the first year, rates for transplantation-free survival were the same for both groups.

The researchers will continue to follow the children in the study over a longer period to further analyze patient outcomes for the two types of shunts.

Beyond survival, patient outcomes include quality-of-life issues. As medical and surgical advances have improved survival rates for children born with heart defects, caregivers have been able to focus on long-term effects of their disease and its treatment. In particular, children with complex congenital heart disease (CHD) are at greater risk of neurodevelopmental problems. As these children reach school age, they tend to have higher rates of academic, behavioral and coordination difficulties compared to peers.

The Children's Hospital of Philadelphia is one of several centers that now have formalized programs to provide ongoing neurodevelopmental care for children with congenital heart disease. As a member of such a program and a leader in ongoing research on neurodevelopmental outcomes in CHD patients, Gaynor concluded, "It will be important to monitor these children as they grow and to not only make sure they are physically doing well. but also hitting developmental milestones."

For more information, visit www.chop.edu.

First Common Gene Found for CHD

Although CHD (congenital heart disease) represents the most common major birth defect, scientists have not previously identified the genes that give rise to it. Now genetics and cardiology researchers, two of them brothers, have discovered a genetic variant on chromosome 5 that strongly raises the risk of CHD.

"This gene, ISL1, plays a key role in regulating early cardiac development, so there is a compelling biological reason for investigating it as a genetic risk factor for CHD," said study leader Peter J. Gruber, MD, PhD, a cardiothoracic surgeon and developmental biologist at The Children's Hospital of Philadelphia. Gruber collaborated with his brother, Stephen B. Gruber, MD, PhD, a geneticist and epidemiologist at the University of Michigan Medical School.

The study appeared online today in the *Journal PLoS ONE.*

Congenital heart disease, said Peter Gruber, is the "Wild West" of genetics, largely unexplored when compared to diseases such as cancer. Researchers have identified genes involved in chromosomal abnormalities and rare genetic syndromes that include heart defects, but no common gene variant had previously been found for non-syndromic complex CHD.

CHD affects at least one in 100 live births. It ranges widely in severity, from tiny holes between heart chambers that close naturally, to life-threatening abnormal structures such as Hypoplastic Left Heart Syndrome that require a series of complicated surgeries.

CHD can affect a variety of different structures in the heart, but the researchers decided to focus on the earliest period of the organ's development. "Instead of assuming separate genes would govern each specific defect, we formed the hypothesis that a common gene variant operates early in the biological pathway of heart formation, thus affecting multiple subtypes of congenital heart disease," said Peter Gruber.

In Peter Gruber's previous research in human cardiac stem cells, he found that a gene called ISL1 was crucial in regulating the development of early cardiac progenitor cells. Suspecting that ISL1 was a likely candidate gene involved in human CHD, he designed a study in collaboration with two genetics teams, one in Philadelphia, the other in Michigan.

At the Children's Hospital of Philadelphia, he worked with Hakon Hakonarson, MD, PhD, director of the Center for Applied Genomics, one of the world's largest centers for pediatric genotyping. Gruber collected DNA samples from 300 children with CHD at the hospital's comprehensive Cardiac Center, and from 2,200 healthy children at the Center for Applied Genomics. Hakonarson's team did the initial genotyping—looking for gene variants (mutations) in the DNA of genes in or near the ISL1 gene. When combined with results from the genetics team at the University of Michigan, the researchers found eight of these alternative spellings in DNA bases (single-nucleotide polymorphisms, or SNPs) raised the risk of CHD.

Stephen Gruber and colleagues at the University of Michigan performed second-stage studies on the initial data, analyzing specific

The Second Fetal Cardiology Symposium

November 19-21, 2010

Scott and Laura Eller Congenital Heart Center
St. Joseph's Hospital and Medical Center, Phoenix, Arizona

The Fetal Heart Center of the Scott and Laura Eller Congenital Heart Center at St. Joseph's Hospital and Medical Center in Phoenix present a three-day symposium for physicians, nurses and sonographers

Guest Speakers:
Norman Silverman, MD
Greggory DeVore, MD
Lisa K. Hornberger, MD
Jack Rychik, MD
Mark Sklansky, MD
Janice L.B. Byrne, MD
Janette Strasburger, MD

Faculty:
Ernerio Alboliras, MD
David Cleveland, MD
John Nigro, MD
Linda Chambliss, MD, MPH
Edward Rhee, MD
Karim Diab, MD
Stephen Pophal, MD

Beth Rumack, RN, NNP
Jonathan Quintana, RDMS, RDCS
Hyehyun Reynolds, RDCS

Individuals interested in attending can register online at **fetalcardio.org** or by calling the ResourceLink at **1-877-602-4111**.

Scott and Laura Eller Congenital Heart Center

Caring for Hearts for a Lifetime

DNA sequences and performing "fine mapping" research—focusing in sharper detail on the gene regions of interest. "It was challenging to analyze how genetic variation contributes to complex congenital heart disease," Stephen Gruber said. "We combined expertise in cardiology, epidemiology, genetics and developmental biology that led to an interesting discovery."

Adding DNA from medical programs in Canada and the Netherlands to the US samples, the researchers studied genes from a total of 1,344 children with CHD and 6,135 healthy controls, and confirmed in replication studies that variants in the ISL1 gene had strong associations with CHD. Within that gene, they found that one SNP raised the risk for white children, and a different SNP increased the risk for African American children.

While the gene findings do not directly affect treatment for children with CHD, Peter Gruber said that better knowledge of the molecular basis of heart disease may provide eventual benefits for the children he sees as a surgeon. "As future studies better define exactly how a mutation leads to a specific type of heart defect, we may be better able to predict how a gene variant affects other organ systems," he added. "We may be better able to understand how a child will respond to surgery, and when or even perhaps how to best perform perioperative, intraoperative or postoperative care. A greater understanding of molecular events in early development brings us that much closer to personalized medicine."

The Leducq Foundation provided funding support for this study. Co-authors with the Grubers and Hakonarson were Kristen N. Stevens - University of Michigan; Cecilia E. Kim, Jennifer Raue, Joseph T. Glessner and Anne Granger - The Children's Hospital of Philadelphia; and collaborators from the Netherlands, Canada and Spain. Peter Gruber is also a member of the Penn Cardiovascular Institute and the Institute for Regenerative Medicine, both at the University of Pennsylvania School of Medicine.

Citation: Stevens KN, Hakonarson H, Kim CE, Doevendans PA, Koeleman BPC, et al. (2010) Common Variation in ISL1 Confers Genetic Susceptibility for Human Congenital Heart Disease. PLoS ONE 5(5): e10855. doi: 10.1371/journal.pone.0010855.

The funders had no role in study design, data collection and analysis, decision to publish, or preparation of the manuscript.

Online version: http://dx.plos.org/10.1371/journal.pone.0010855.

TINY HEARTS INSPIRED HYBRID LABS WITH ACCESS FOR BIG TEAMS.

Fixing a heart from birth through adulthood takes big teams working together. So we examined the needs of leading clinicians when designing our hybrid solutions. The result: our Infinix™-i with 5-axis positioners and low profile detectors, stays out of the way, but right where needed, providing the best possible access to patients. To lead, you must first listen.

medical.toshiba.com

TOSHIBA
Leading Innovation >>>