PEDIATRIC CARDIOLOGY TODAY

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VOLUME 2, ISSUE 2 WWW.PEDIATRICCARDIOLOGYTODAY.COM FEBRUARY 2004

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USE OF THE EXCIMER LASER IN THE INITIAL MANAGEMENT OF PULMONARY VALVE ATRESIA WITH INTACT VENTRICULAR SEPTUM

By Michael C. Slack, MD

Introduction

Lasers (an acronym for Light Amplification by Stimulated Emission of Radiation) from various light sources have been used in medicine for over 30 years. In cardiovascular medicine, in the 1980s, various lasers including those from Argon and YAG sources found limited use. The excimer laser (an acronym for excited dimer), was found useful in the angioplasty of clogged saphenous vein grafts.(1) More recently, the excimer laser has been refined for use in various cardiovascular indications such as coronary artery revascularization, treatment of In-stent restenosis, thrombosed vessel revascularization, peripheral vascular disease, transmyocardial revascularization, and pacemaker lead removal. Until recently, the use of lasers in the management of congenital heart disease was limited primarily to pacemaker lead revision procedures.

> ***"Further refinements in the design of the excimer laser catheters, including significantly smaller catheter size for the coronary and peripheral vascular markets, have made this technology potentially useful to the pediatric interventional cardiologist."***

Further refinements in the design of the excimer laser catheters, including significantly smaller catheter size for the coronary and peripheral vascular markets, have made this technology potentially useful to the pediatric interventional cardiologist. In this report, the use of the latest extremely small profile excimer laser catheter is described in order to produce a controlled discrete opening in the imperforate membrane of an atretic pulmonary valve as a method of performing successful balloon valvuloplasty in the initial palliative management of this uncommon condition. Additionally, we will present some of the basics of the excimer laser and information regarding the effective use of the recently available and FDA approved small diameter concentric "Extreme" laser catheters.

Case Report

A 36 week gestation male infant, birth weight 2.8 kg, was evaluated at one day of age for circumoral cyanosis and a heart murmur. His clinical evaluation, including a complete transthoracic echocardiogram revealed pulmonary valve atresia with intact ventricular septum. The atretic pulmonary valve morphologically consisted of a completely intact membrane without evidence of antegrade flow by color Doppler. There was also

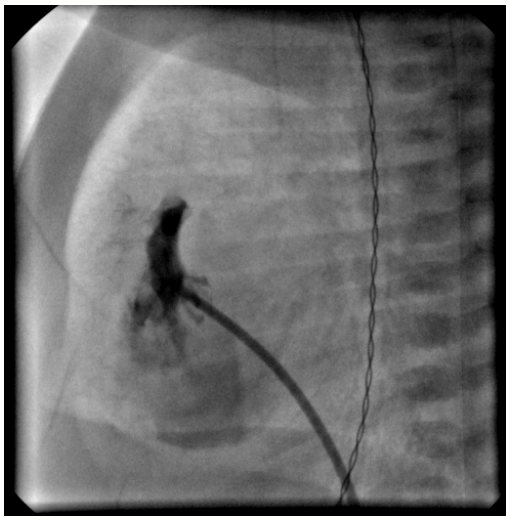

Figure 1. Lateral view of RVOT angiogram showing the completely intact atretic pulmonary valve. Note: Visible rudimentary valve leaflets below the intact membrane.

Figure 2a. (see text for description)

Figure 2b. (see text for description)

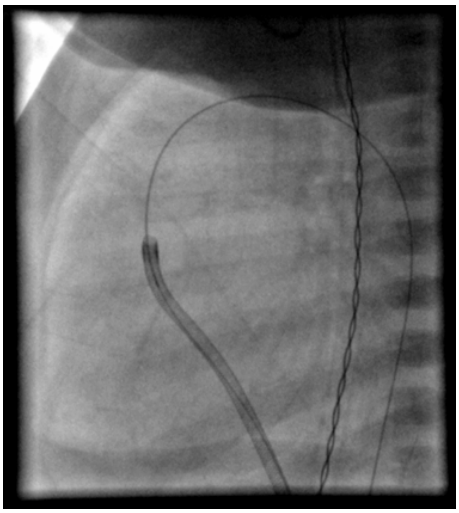

Figure 2c. (see text for description)

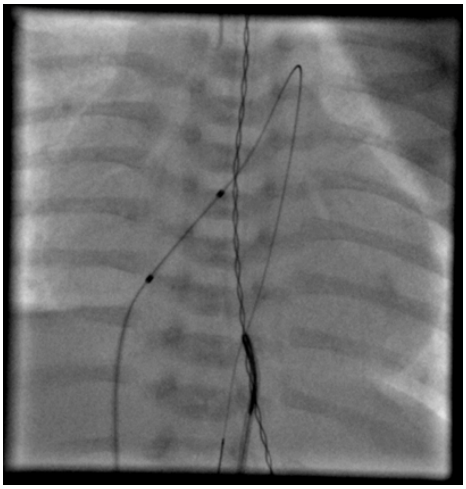

Figure 2d. (see text for description)

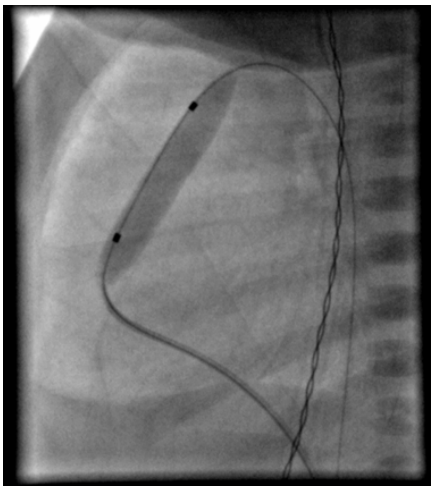

Figure 2e. (see text for description)

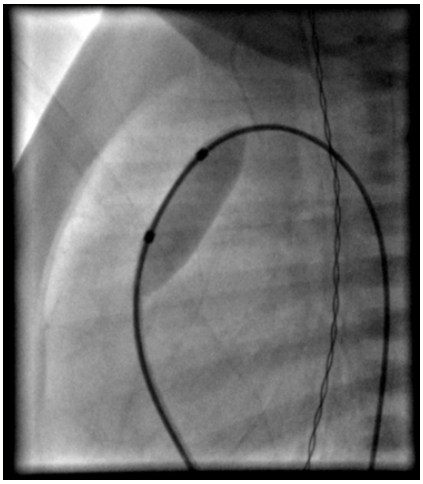

Figure 2f. (see text for description)

a mild to moderate severe Ebstein's malformation of the tricuspid valve with mild tricuspid regurgitation (TR). There was a stretched patent foramen ovale and a large patent ductus arteriosus (PDA). A continuous infusion of prostaglandin (PGE 1) was immediately instituted.

At about a week of age, informed consent was obtained and the infant was taken to the cardiac catheterization laboratory for pulmonary valve perforation and balloon valvuloplasty. Initial diagnostic catheterization confirmed the diagnosis and angiography in the right ventricular outflow tract (RVOT) demonstrated a completely intact pulmonary valve membrane (Figure 1). The CVX-300 Excimer Laser System (Spectranetics Corporation, Colorado Springs, CO), previously warmed up for 20 minutes, was then employed. A 6 French FR4 (renal) guide catheter (Boston Scientific Corp., Natick, MA)

"Importantly, there are serious safety concerns and potential consequences from the improper use of the laser equipment..."

"Until recently, the use of lasers in the management of congenital heart disease was limited primarily to pacemaker lead revision procedures."

was advanced over a .035 angled Terumo "glide" wire (Boston Scientific Corp., Natick, MA) into the right ventricular outflow tract. Using both angiographic and transesophageal echocardiographic imaging, the FR4 guide was manipulated into optimal position in the RVOT. A Point 9™ Extreme (over-the-wire) Excimer Laser catheter (Spectranetics Corporation, Colorado Springs, CO) was advanced over a 0.014 BMW coronary wire (Guidant Corp., Santa Clara, CA) through the prepositioned guide. The wire and laser catheter were advanced taking care to maintain the floppy tip of the wire beyond the tip of the laser catheter at all times. The floppy tip of the wire was allowed to form a "u" shaped loop on the underside of the valve membrane while the laser catheter was advanced and centered on the valve (Figure 2a). Once positioning was confirmed by TEE and angiographic roadmap, the wire was retracted to the tip of the laser catheter and laser energy was applied (45 mJ/mm(2);15 kV;25 Hz). The laser catheter was noted to cross the valve within several seconds and the wire was immediately advanced into the main pulmonary artery (Figure 2b), down the PDA and into the descending thoracic aorta (Figure 2c). The wire was snared and brought out of a 4 French arterial sheath (Figure 2c). A 6mm x 2 cm Symmetry balloon (Boston Scientific Corp., Natick, MA) was then advanced over the BMW wire and advanced across the valve using forward pressure and firm pulling traction from the arterial side of the wire loop (Figure 2d). Two inflations to 8 atmospheres were performed (Figure 2e). The wire was then up-sized to a .035 wire and two additional inflations were performed using 8mm x 1.5 cm Ultra Thin Diamond balloon (Boston Scientific Corp., Natick, MA) (Figure 2f). Anatomic separation of the valve leaflets with good forward flow through the valve was visible angiographically (Figure 3). No change in the degree of tricuspid regurgitation (2+) was seen however, negative washout from competitive PDA flow was noted.

"One drawback is the warm-up time of nearly 20 minutes for the console requiring advanced planning for use of the system."

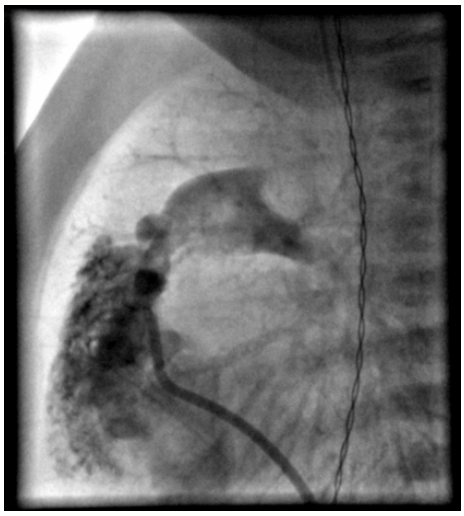

Figure 3. Lateral right ventricular angiogram following successful laser perforation and balloon valvuloplasty with 6mm then 8mm diameter balloons. Note the valve leaflets now separated and in the open position (arrow).

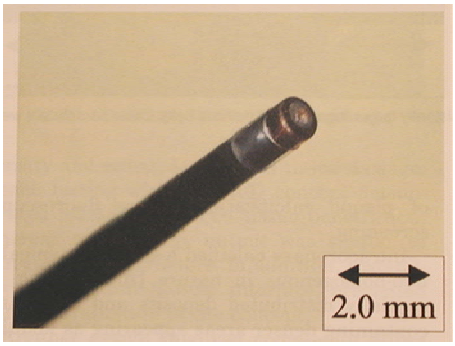

Figure 4. a-side view of the 0.9 Extreme excimer laser catheter tip. Spectranetics, with permission

Initially the infant did well with pulse oximetry averaging in the low to mid 80s. Over the next week, with the PDA now closed, the infant had periods of significant desaturation. Echocardiography demonstrated a velocity of 3.2m/s across the pulmonary valve with mild pulmonary insufficiency and 2+ TR. Additionally, significant right to left shunting was noted at the atrial level. Repeat balloon pulmonary valvuloplasty was performed with a 10mm x 2 cm with reduction of the valve gradient from 34mmHg to 6mmHg. A follow-up angiogram demonstrated improved valve leaflet excursion with excellent forward flow through the valve. Pulmonary insufficiency increased from mild

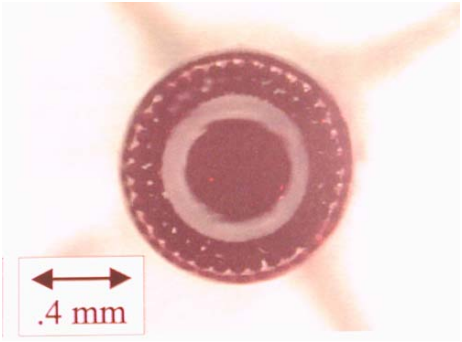

Figure 4b-detail of catheter tip showing the concentric ring of fiber bundles surrounding the wire lumen. Spectranetics, with permission.

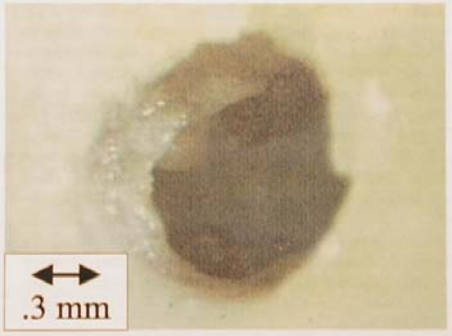

Figure 5. Hole lased in an ex vivo human tissue sample with the 0.9 mm Extreme excimer laser catheter (33sec;80mJ/mm2 & 60Hz). Note: well circumscribed discrete edges. Spectranetics. with permission.

to 1.5+ and TR remained unchanged. The infant improved markedly and was discharged from the hospital by three and a half weeks of age. At six month follow-up, his pulse oximetry is averaging 91% with significantly improved right ventricular growth. There is a small residual atrial septal defect with predominately left to right shunting.

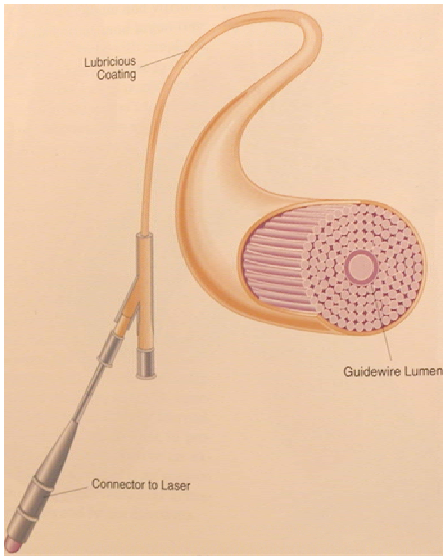

Figure 6. From: Litvack F, Eigler NL, Forrester JS. Excimer Laser Coronary Angioplasty. Science & Medicine, 1996;3(1):42-51; with permission.

Discussion

Valve perforation in the management of intact membranous type of pulmonary valve atresia is a technique which has been in a state of continual albeit slow evolution. The combination of extremely low procedural volumes with no specifically designed tools, have provided the interventional cardiologist with the opportunity to be creative. Stiff-end wire perforation(2), radiofrequency ablation, catheter-assisted perforation,(3)(4) and laser catheter-assisted perforation,(5)(6) have been described. Reports of the use of excimer lasers in congenital interventional procedures have been few due in large part to the limited stearability and requirement for large sheaths. Recently, however, due to the introduction of the Extreme Point 9™ line of laser catheters from Spectranetics, the potential utility of this versatile laser catheter has been realized.(7)

The excimer laser uses light in the ultraviolet wavelength (308 nm) produced as an excited dimer from xenon chloride gas. The resulting laser light ablates tissue by delivering extremely short bursts of very high light energy that result in disruption of the carbon to carbon double bonds, a process called "photo molecular dissociation." The depth of the lesion can be manipulated but is usually no more than 100µ deep per laser application. Ninety-five percent of the particles formed from tissue ablation are < 5 microns in size. The catheters consist of fiber optic bundles surrounding a central wire lumen running the entire length of the catheter (Figure 4). In vitro testing confirms that the catheter creates an extremely reproducible and discrete lesion in tissue (Figure 5). We have found the Extreme Point 9™ laser catheter very user friendly and extremely trackable (Figure 6). The CVX-300 Generation 4 Excimer Laser System from Spectranetics Corporation is FDA approved for percutaneous revascularization in patient with coronary artery occlusive and lower limb vascular disease. One drawback is the warm-up time of nearly 20 minutes for the console requiring advanced planning for use of the system. Importantly, there are serious safety concerns and potential consequences from the improper use of the laser equipment, therefore all potential users must complete a formal training course sponsored by Spectranetics Corporation prior to clinical usage (contact your local salesperson to arrange training).

The recommended CPT Code by Spectranetics is 35480

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MEDICAL CONFERENCES

ACC '04 (Annual Scientific Session)
March 7-10, 2004, New Orleans, LA
www.acc.org

Electrophysiology in the Young and Adults: A New Frontier
March 23-25, Riyadh, Saudi Arabia
www.kfshrc.edu.sa

29th Annual Scientific Meeting - 2004 SIR (Society of Interventional Radiology) "The World of Intervention"
March 25-30, 2004, Phoenix, AZ
www.sirweb.org

39th Annual Meeting - Association for European Paediatric Cardiology
May 19-22, 2004 - Munich, Germany
www.i-plan.de/aepc

14th World Congress in Cardiac Electrophysiology and Cardiac Techniques
June 16-19, 2004, Nice, France
www.cardiostim.fr

81st CPS Annual Conference (Canadian Paediatric Society)
June 16-20, 2004, Montreal, Canada
www.cps.ca/english/index.htm

Society of Nuclear Medicine 51st Annual Meeting
June 19-23, 2004, Philadelphia, PA
interactive.snm.org

International Society for Minimally Invasive Cardiac Surgery (ISMICS) Annual Scientific Meeting
June 23-29, London, UK
www.ismics.org

The 2004 "Specialty Review in Pediatric Cardiology" Course
July 12-15, 2004, Chicago, IL
www.uic.edu/depts/ci/pcard04

THE PAPERLESS OFFICE

By Warren H. Toews, MD

Compared to the airline and banking industries, healthcare lags dramatically behind in information management. And yet, few of us would dispute the necessity for readily accessible, complete medical records in the hospital and in our offices. In addition, the economic pressures of practice today require greater efficiencies.

Fewer than 10% of physicians in the U.S. currently use any form of Electronic Medical Record (EMR) generation or storage, and fewer than 2% of practices are completely paperless. Old habits die hard.

The most commonly listed reason for using an EMR is access to patient records. Traditionally, charts are kept in file cabinets or rooms. They require time to pull, and finding charts can occasionally be challenging. Other reasons to use an EMR include decreased dictation time and transcription expense, improved documentation and E & M Coding, better HIPAA compliance, better handling of prescriptions, and easier communication with referring physicians.

EMR's come in 3 basic varieties. The most basic type is a "virtual transcriptionist", typically using either a menu-driven keyboard or voice-activated data entry system, still requiring that the report be manually printed, mailed, copied and filed.

For paperless offices, some kind of data storage system is required. There are Application Service Providers (ASP) that maintain records on their website, and physicians enter data over the Internet. Although these systems are fairly inexpensive, there are concerns about patient confidentiality, fragility of the Internet connection, and the fate of the records if the ASP goes out of business. Perhaps the biggest disadvantage is that most of these systems were made for primary care practices, and require quite a lot of adaptation for cardiology practices.

There are also "stand alone" systems. We chose to use such a Cardiology-specific product with the file server located in our office. It is much faster than most Internet connections, has the necessary firewalls, and the data is appropriately backed up.

After looking at a number of products in 2000, we chose AllMeds for Cardiology, one of the specialty-specific products from AllMeds*. Although originally designed for adult cardiology, it suits our needs quite well, and is easily modified for our purposes through the use of standard notes and stored outlines.

> ***"Fewer than 10% of physicians in the US currently use any form of Electronic Medical Record (EMR) generation or storage…"***

Our system is a searchable database, so that any data entered in any previous patient visit can be "pulled forward" to the current entry, saving time, and making for more complete records. We only make appropriate changes in the data. Our system will perform an E & M Coding check automatically, allowing us to be sure that our billing is appropriate. Since all of the records are electronically signed off, they cannot be altered at a later time. It is password protected. Our office employees have access only to those parts of the record that are necessary for their jobs.

Currently, in our practice, access is immediate, typically requiring seconds to pull up any of our nearly 10,000 patient records on the computer. Files are available from either of our two metro-Denver offices, as well as from our homes (great when on call), and from any of our seven remote outreach sites (via the Internet). Phone calls from other physicians or from patients are easily and quickly dealt with. We can search for records by any demographic identifier, by referring physician, by diagnosis, etc.

AllMeds for Cardiology has a series of templates for HPI, PMFSH, ROS, and Physical Exam. Each template has a "drop-down" menu so that the entire template can, if desired, be filled out by mouse clicks. I typically use few of those, often preferring to use one of a series of standard notes and stored outlines I have composed. For example, I have notes for new patients referred for murmurs, one for babies and one for older children. I simply pull up the note, use my keyboard to make any necessary changes, and then enter it in the appropriate place. I do the same for follow-up patients. The record automatically enters the reason for follow-up (i.e., diagnosis), and I use a standard follow-up note that includes the date of the last visit, modifying it if necessary for changes in the history. After the first follow-up visit, if the symptom status is unchanged, all I need do is change the date of the previous visit and make any changes in the history. Prior to implementing the AllMeds EMR, reports required a minimum of 2 working days to process--from dictation to mailing. Now, we generate reports from communications templates in our

system and fax them to the referring physicians, often directly from the exam room, before the patient leaves. I also provide the parents with a copy of my full report as they leave the office. This has proved invaluable when patients have to visit an ER when on vacation, etc.

I use stored outlines for HPI, Past, Family and Social History, ROS and Physical Exam. These populate the template with negatives and normals, and I just alter those that are incorrect. After the first visit, I only enter changes. Many practices employ a scribe, usually a PA, MA, or RN, who performs the computer data entry while the physician takes the history and does the exam. Since the scribe works with the same physician every day, he or she knows what the physician wants in the report. In some practices, this technique has increased patient throughput by up to 30%.

Our prescription-writing module not only checks for allergies and alerts us to possible drug interactions, but also prints the prescriptions, or faxes them based on a built-in pharmacy database. There are never questions about handwriting. We also can enter any number of Patient Information Sheets that we can access, display to the patient, and print immediately. Any printed data received from other sources is simply scanned in to the patient file. Our previous file clerk is now a scanning clerk.

Transcription previously cost our 3-physician practice about $4500/mo. Comparatively, our cost for AllMeds' EMR is about $3,000 per month. This includes the software license, support, training and maintenance, along with a 3-year equipment lease. After the first year, we have paid about $1000/month for software support and upgrades. We incurred some extra expenses with the recent opening of our second office in Denver. The office required quite a bit of IT work to get high speed access to the server from the second office, and we needed to upgrade some of our hardware. We have experienced a small savings compared to using transcription after factoring in costs associated with startup and expansion. A larger practice may experience a greater financial benefit.

AllMeds has a good product. During the three years of our usage they have demonstrated a commitment to support and service. Furthermore, the company has been open to suggestions for software changes. AllMeds for Cardiology has helped improve the quality of our patient records, improved workflow, and most importantly, given us immediate and complete access to our patient records.

For comments to this article, send email to: FEBWT@PediatricCardiologyToday.com

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Preliminary EMR Checklist

- ☑ Access to records
- ☑ Compile records
- ☑ Legible records
- ☑ Searchable database
- ☑ Prescription writing
- ☑ Cost savings

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Western Cardiology Associates
Division for Fetal, Pediatric, and Adult Congenital Heart Disease

Emergency Medical Records

Selected List of EMR Companies

***AllMeds**
www.allmeds.com

Amicore, Inc.
www.amicore.com

ComChart EMR
www.comchart.com

DocuMed
www.documed.com

e-MDs
www.e-mds.com

GEMMS
www.gemmsnet.com

MedInfomatix
www.medinformatix.com

MEDCOM
www.emirj.com

NextGen EMR
www.nextgen.com

PowerMed EMR
www.powermed.com

SynaMed
www.synamed.com

TCAOS
www.tcaos.com

EMR Information Resources

EMR Comparisons
www.elmr-electronic-medical-records-emr.com

Healthcare Informatics Online (see Spotlights)
www.healthcare-informatics.com

CTS Software Selection
www.ctsguides.com/medical-method.asp

CONGENITAL HEART BLOCK IN INFANTS BORN TO MOTHERS WITH SLE: MATERNAL-INFANT AUTOANTIBODY MARKERS AND MANAGEMENT STRATEGY

By Z. Zain, MBBS; A. A. Majid; A. Omar, MBBS; C. S. Khuan, MBBS; and J. Hassan, MBBS

Introduction

The association between maternal systemic lupus erythromatosus (SLE) and adverse fetal outcome due to the transplacental transfer of autoantibodies has been widely reported. Congenital heart block is one of the manifestations of neonatal lupus syndrome and is a recognized cause of morbidity and mortality in infants born to mothers with SLE.

Autoantibodies to SSA/Ro and SSB/La ribonucleoproteins have been demonstrated almost universally in the maternal circulation when isolated congenital heart block is identified [1]. These are soluble nuclear (SSA/Ro) and soluble cytoplasmic (SSB/La) autoantibodies which are directed against cellular ribonucleoprotein complexes predominantly found in patients with sicca syndrome or SLE and is readily identified by the double immunodiffuson method [2].

The presence of these autoantibodies is strongly associated with neonatal lupus, a disorder considered as a model of passively acquired autoimmunity [3]. Complete heart block which is a potentially severe and permanent manifestation of neonatal lupus appearing after the first trimester of pregnancy is an example of this model. The mechanism of disease is dependent on the placental transport of maternal antibodies (Anti-SSA/SSB) capable of causing specific myocardial inflammation that permanently damages the conduction system of the developing fetal heart [4].

We describe the management and outcome of congenital heart block in infants of mothers with SLE and the association of maternal-infant lupus autoantibody markers.

Patients and Methods

All study infants were born in the single centre in which the study was reported. All infants were identified during fetal ultrasound screening for pregnant mothers with SLE between August 1999 till March 2002.

Connective tissue disease screening including lupus autoantibodies in mothers were examined during antenatal check and the extracted nuclear antigen (ENA) screen was performed for positive serum.

Fetal echocardiography was performed periodically and standard views were obtained to identify structure and intracardiac connections. Fetal M-mode tracings were recorded to determine atrioventricular dissociation and estimate left ventricular contractility.

All pregnancies were seen up till term gestation and delivery was planned with the cardiologist and cardiac surgeon on standby.

Infants born were seen immediately by the cardiologists for cardiac assessment. Blood samples were obtained for ANA and specific nuclear antigens.

No	Maternal ANA	Maternal anti-Ro/ anti- La	Baby anti-Ro/ anti- La	Type of Heart Block
1	Positive	Pos / Pos	Pos / Pos	Complete Heart Block
2	Positive	Pos / Neg	Pos / Pos	Complete Heart Block
3	Positive	Neg / Neg	Neg / Neg	Complete Heart Block
4	Positive	Pos / Pos	Pos / Pos	Complete Heart Block
5	Positive	Pos / Pos	Pos / Pos	Complete Heart Block
6	Positive	Pos / Pos	Pos / Pos	Complete Heart Block
7	Positive	Pos / Pos	Pos / Pos	Complete Heart Block
8	Positive	Pos / Pos	Pos / Pos	Junctional Heart Block

Table 1. Maternal-infant lupus autoantibody profile

Results

There were eight patients in this study. There were seven girls and one boy. All patients were diagnosed at the mean gestational age of 28 weeks. Fetal echocardiogram showed normal intra-cardiac structures in all patients with bradycardia,

dilated and moderate to poorly contracting ventricles in seven patients. Fetal cardiac M-mode recorded complete AV dissociation in these seven patients with mild to moderate degree of pericardial effusion. No treatment in-utero was given.

In one patient, the fetal bradycardia was detected during routine ultrasound on the mother and investigations revealed that she was positive for SLE autoantibodies including anti-Ro/SSA despite not being having active disease. Only one of the mothers was in active disease at the time of delivery and she and the baby succumbed to haematological complications of SLE.

The mean gestational age was 36 ± 4 weeks. Six were delivered by Caesarean section, two vaginally. Mean birth weight was 2.65 ± 0.4 Kg. All patients had surface electrocardiogram recording and transthoracic echocardiogram performed.

Seven patients (87.5%) had complete AV block and one had a slow fixed junctional rhythm. The mean intrinsic heart rate was 46 ± 13 beats per minute.

The mean left ventricular internal diastolic diameter (LVIDD) was 2.3 ± 0.6 cm (mean z-score = 1.9 ± 0.4) and the mean left ventricular ejection fraction was 36 ± 7%. Other findings were pericardial effusion in the same seven patients (with complete AV block) and valvular pulmonary stenosis in one patient.

Six (75%) of the patients underwent urgent thoracotomy for epicardial wire insertion and temporary ventricular pacing within 12 hours of delivery. Two newborns had immediate general anesthesia and epicardial wire insertion performed with the umbilical cord unclamped and placenta not separated until the wires were secured and external pacemaker connected. Patients were then managed in the hospital and permanent pacemaker insertion was performed electively. The median time of permanent pacemaker implantation (n=6) was 30 days (range 18-180 days).

The implantable pacemakers used in all patients were from St Jude's Medical (Models Microny™ K SR, Microny™ SR+ or Microny™ II SR+) all of which are designed to be used in small patients who needs high base rate pacing (12.8grams, 5.9cc and 6mm thin).

One patient with complete AV block had complete spontaneous recovery to sinus rhythm without treatment. She remains well on follow-up. Two patients died. One patient with a fixed junctional block (rate 60 per minute with isoprenaline) died of hematological complications related to SLE soon after birth. Another patient died after pacemaker insertion due to respiratory complications at the age of 2 months.

Lupus Autoantibody Profile

All mothers (n = 8, 100%), had ANA positive. Seven (87.5%) of the babies had evidence of transplacental transfer of lupus autoantibodies in which 6 (75%) mother-baby pairs were positive for the anti-SSA/Ro and anti-SSB/La (Table 1). Two babies were strongly positive for ANA (> 1:1000), 3 had in addition anti-sm/RNP, anti-cardiolipin 7 and anti-Jo1 positive respectively.

Period of follow-up is between 1-3 years. All patients with pacemakers are thriving well with satisfactory normal left ventricular dimensions and function on echocardiogram. None of the patients are on any cardiac medications. The outcome of patients in this study is shown in figure 1.

Discussion

Neonatal lupus is characterized by cutaneous lesions, heart block or both [2]. The type of in-utero heart blocks reported range from partial to complete atrioventricular block and is due to fibrosis of the conduction system. Ante-natal detection has allowed peripartum management including use of beta-agonists and steroids but the mainstay of treatment remains the insertion of single chamber ventricular pacemaker.

USEFUL WEBSITES
Associations and Societies

HighWire - Library of the Sciences and Medicine
highwire.stanford.edu

Heart Foundation of South Africa
www.heartfoundation.co.za

The Hypertrophic Cardiomyopathy Association (HCMA)
www.4hcm.org/home.php

Indian Society of Vascular and Interventional Radiology (ISVIR)
www.isvir.org

International Academy of Cardiology
www.cardiologyonline.com

The International Society for Minimally Invasive Cardiac Surgery (ISMICS)
www.ismics.org

The Mid-America Interventional Radiological Society
www.mirs.org

The National Association of Children's Hospitals and Related Institutions (NACHRI)
www.childrenshospitals.net

National Center for Early Defibrillation
http://198.65.239.253

National Electrical Manufacturers Association (NEMA)
www.nema.org

National Medical Association (NMA)
www.nmanet.org

NASPE-Heart Rhythm Society
www.naspe.org

Mothers with SLE are at risk to bear children with congenital heart block. The overall risk of giving birth to an infant with congenital heart block, among women with probable or definite SLE was 1:60 but when the woman had anti-SSA/Ro, the risk was 1:20 [5].

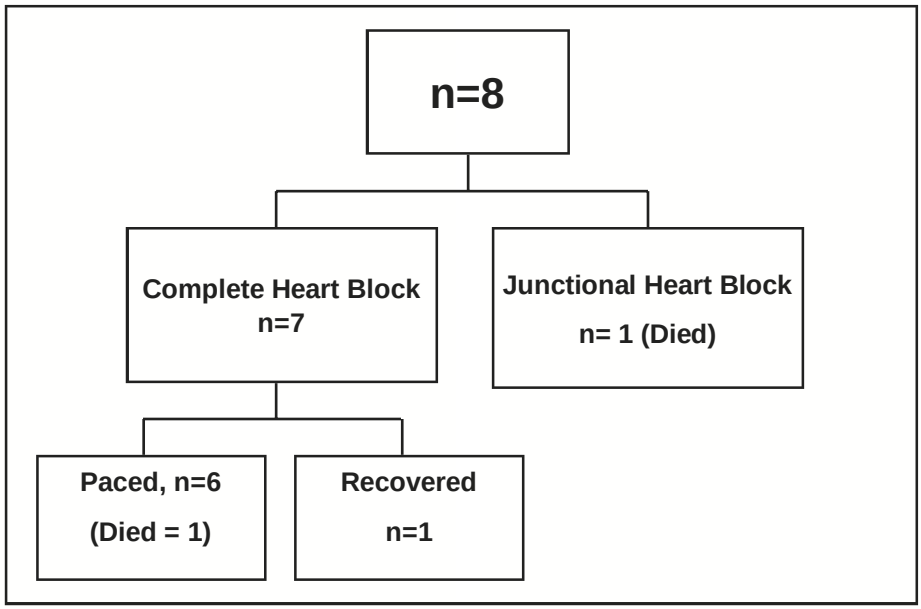

Figure 1. Outcome of patients with congenital heart block

In this study, all infants were detected during routine ultrasound check for women with SLE who were pregnant. These patients were then carefully followed up till term gestation with pre-existing management plans in order for the infant with congenital heart block except for one. In one patient, the mother had clinical features of SLE including hematological disease and died post-partum secondary to disseminated intravascular coagulation (DIVC). The baby had a fixed junctional type of heart block of 60 beats per minute despite intravenous infusion of isoprenaline and succumbed rapidly due to severe hematological abnormalities. No pacemaker insertion was attempted.

All babies with decompensated complete AV block (n=6) had an external temporary pacemaker with epicardial wires inserted to the right ventricle by thoracotomy soon after birth. Cardiac decompensation was determined clinically as the presence of signs of poor cardiac output. Two newborns had their procedure done on a side table to the mothers with the umbilical cord unclamped with the placenta intact in the uterus as soon as the baby was delivered by caesarean section. The patent flow across the umbilical vessels provided adequate volume and stimulation and both procedures were completed without any complications.

Permanent subcutaneous implantable pacemakers (single chamber) with bipolar steroid eluding epicardial leads were electively inserted in six patients. The optimal time for permanent pacemaker insertion was determined by the weight of the baby and this was achieved by establishing adequate nutrition and treating concurrent infections.

Previous management experience in our institution before this study for a small number of babies with similar history were not satisfactory. Transvenous pacemaker wire was inserted either by the femoral vein or umbilical vein route while awaiting permanent pacemaker insertion which was complicated by infection at the insertion site or recurrent dislodgment of the lead wire.

Patients weighing or had achieved weights of 3.2 Kg and above were scheduled for permanent pacemaker insertion. Pacemaker implantation was done in a subcutaneous pocket created in the subcostal region in all patients and epicardial leads secured to the epicardium adjacent to the right ventricle and allowing excess wire to allow for growth and movement. We found this strategy of staged approach to have good outcome albeit longer stay for the patient in hospital.

The experience with the existing patients in this study has allowed us to organize a multi-disciplinary approach in the management of these infants beginning from ante-natal period in which pregnant mothers are counseled by the rheumatologists and obstetricians. Upon detection of any cardiac abnormalities particularly heart block, the pediatric cardiologists and cardiac surgeons are then involved in the planning of the post-natal management of these infants.

Perinatal treatment has not been proven to be useful to improve heart rate or ventricular contractility except for experiences in small numbers of patients [14-17] and therefore not practiced in our center. Steroids are used, however, to expedite lung maturity if necessary.

Permanent pacemaker insertion is the definitive treatment for babies with congenital complete AV block and the ideal situation would be to be able to manage them through perinatal period and achieve a safe delivery.

Miniaturized permanent pacemakers in this current era has allowed early insertion and eliminated the issue of the infant's size and weight for selected centers.

"Congenital heart block is one of the manifestations of neonatal lupus syndrome and is a recognized cause of morbidity and mortality in infants born to mothers with SLE."

We have adopted the staged approach for pacemaker insertion in our infants in view of our limitations and this strategy has shown to have good outcome.

Conclusion

Congenital heart block is a rare but serious fetal adverse disease due to maternal-fetal transmission of anti-SSA/Ro in pregnant mothers with SLE. Anticipation of this condition and early detection by fetal echocardiography allows an organized multi-disciplinary management with a staged approach which promises favorable results.

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~See author for additional references~

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~PCT~

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Pacemaker Information

Selected Pacemaker Companies

St. Jude Medical, Inc.
✆ (+1-651) 483-2000
www.sjm.com

Medtronic
✆ (+1-763) 514-4000 or 574-4000
www.medtronic.com

BIOTRONIK GmbH & Co.
✆ (+49 30) 6 89 05 –0
www.biotronik.com

Guidant Corporation
✆ (+1-317) 971-2000
www.guidant.com

Selected Resources

American Heart Association
www.americanheart.org

American College of Cardiology
www.acc.org

theheart.org
www.theheart.org

U.S. Food & Drug Administration (FDA)
www.fda.gov

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