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Early Post-Operative Risk Factors After Fontan: Experience from The Rajaie Heart Center, Iran

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Abstract Objectives

The post-operative morbidity and mortality of the Fontan operation are still a matter of concern. The purpose of this report is to assess suspected predictors of unstable hemodynamics and death after the Fontan procedure. Methods and Materials: From 1994 to 2007, we performed a case series study on 80 patients with mean age of 8.5±4 years who had undergone total cavopulmonary connection. Association between the clinical outcomes and suspected risk factors were investigated by student's test (for interval data) and chi square or Fisher's exact test and also poisson regression models. P value < 0.05 considered statistically significant. Results: Unstable hemodynamics and death were evaluated for their association with suspected predisposing variables including; age, weight, sex, pump time, associated heart anomalies with single ventricle, pulmonary artery pressure, previous palliation and different types of Fontan surgery. Statistical analysis showed significant association of death with pump time (P=0.002), pulmonary artery pressure (P<0.001), ventricular morphology (P=0.049) and types of Fontan operation (P=0.035). Conclusion: Decreasing the pump time through using trouble-free kinds of Fontan procedure such as extra-cardiac conduit might decrease postoperative morbidity and mortality. Likewise, considering ventricular morphology beside other prominent risk factors such as mean pulmonary artery pressure and pulmonary arterial size might diminish the undesirable outcomes early after operation.

Introduction

The Fontan operation was reported for the palliation of single ventricle anatomy in 1971.[1,2] Although Fontan's procedure has improved considerably with time, perioperative mortality still occurs, attributed to low pulmonary vascular bed compliance, ventricular dysfunction, stroke, arrhythmia, thromboembolism, and multi-organ dysfunction. Selection of appropriate patients with lower risk factors for operation was performed according to parameters such as pulmonary pressure, size of pulmonary artery branches, atriovenricular valve function and ejection fraction. Some investigators have reported long duration of cardiopulmonary bypass or aortic cross-clamping as the risk factors. [2-4]

Although previous postoperative mortality of 25%-30% has been improved with time, however, most large centers still report mortality rates of 2–7%.[2-4]

Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

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1. Hoffman et al, Anesthesiology 2008; 109 A7 2. Dent et al, J Thorac Cardiovasc Surg. 2006 Jan;131(1):190-7
3. Andropoulos et al, Anesthesiology 2007; 107: A209

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We attempted to evaluate the association of presumed risk factors with hemodynamic instability and mortality.

Materials and Methods

In this study, we evaluated patients with single ventricle between 2 and 24 years old who had undergone Fontan operations during the period 1994 - 2007 in The Rajaie Heart Center in Tehran, the main referral center for pediatric heart disease in Iran. This study was approved by Rajaie Heart Center research committee which is licensed via Iranian health minister. No informed consent was included due to the retrospective structure of our study.

Post-operative death and hemodynamic status were considered outcomes which would be compared statistically with suspected preoperative or perioperative risk factors.

The early post-operative period was defined as the period after termination of Fontan operation until the discharge from ICU.

Anatomically, patients included those with different types of single ventricle anatomy with double, single or common inlet ventricle, However, no patient with Hypoplastic Left Heart Syndrome was treated with the Fontan operation.

Catheterization was performed for all patients at least one time before Fontan operation and in 88%, it was also performed before the primary palliation.

Pulmonary artery pressure measurement was measured by passing catheters through systemic to pulmonary shunts, patent arterial duct, collaterals (75%) or by passing via pulmonary valve (12%). In the few remaining cases it was evaluated by means of pulmonary venous wedge pressure. Increased mean pulmonary artery pressure was considered to be as a crucial risk factor for Fontan operation. Mean pulmonary artery pressures equal to or lower than 15 mm Hg was considered optimal; pressures between 15 and 19 mm Hg were considered high-risk; and pressures higher than 20 mm Hg excluded patients from Fontan surgery. Pulmonary artery branch diameter Z values within 2 Standard Deviations (SD), smaller than between 2 and greater than 3 SD, and smaller than 3 SD were considered respectively to be acceptable, relatively small, or severely hypoplastic.

Echocardiographic evaluation was performed before the operation at least three times under the supervision of a senior pediatric cardiologist. Likewise, echocardiography and contrast echo were carried out after Fontan procedure for anatomic and hemodynamic measurements.

Four types of cavopulmonary connection were classified including; atriopulmonary connection(16.3%), intracardiac lateral tunnel(46.3%), extra-cardiac conduit(26.3%) and bilateral Glenn(11.3%).

Before 1993 atriopulmonary connection was the dominant type of Fontan operation. Thereafter, it declined remarkably as it was no longer performed after 1999, and other types of Fontan procedure including lateral tunnel and extra-cardiac conduit were the main types of operation.

No patients had anomalous pulmonary venous connections.

All types of Fontan operation were started with median sternotomy. Cardiopulmonary bypass was performed by using bicaval cannulation through the superior and inferior caval veins. Cold crystalloid solution used as cardioplegia intermittently. Aortic cross clamp was performed for different types of Fontan procedure with mean time of 50 minutes. Circulatory arrest was not used in our series. Systemic hypothermia was controlled at about 25° C.

In lateral tunnel procedure after anastomosis of incised distal superior caval vein to superior portion of right pulmonary artery a tunnel baffle made of a tube of polytetrafluoroethylene provided a pathway from inferior caval vein toward pulmonary artery leaving a small aperture.

Extra-cardiac conduit was started by anastomosis of the superior caval vein to right pulmonary artery and opening of the inferior surface of right pulmonary artery in order to make the anastomosis to an appropriate polytetrafluoroethylene tube graft connected to the inferior vena cava.

Bilateral cavopulmonary connection was performed in those patients with suitable size right and left superior caval vein, and with azygos continuity and interrupted inferior caval vein.

Atriopulmonary connection was performed by direct anastomosis of main pulmonary artery or right pulmonary artery to the right atrium usually through its appendage.

Early after Fontan procedure all patients were hemodynamically monitored through central venous access and arterial line in the pediatric intensive care unit. Likewise, patients were controlled for cardiac rhythm and oxygen saturation. In addition to pulse oxymetery, arterial blood gas was measured every two hours for the first 24 hours followed by wider intervals during the next days. Hemodynamic instability was determined by means of all clinical and paraclinical measurements.

Continuous intravenous heparin was started as soon as possible depending on the patient's blood coagulation status and quality of chest tube drainage followed by warfarin administration with gradual discontinuation of heparin. Fontan pathway and fenestration flow were assessed at patient's admission to the ICU, and, thereafter serially by means of echocardiography and contrast echocardiography. We intended to keep the mean pulmonary pressure bellow 16 mm Hg as evaluated by pulmonary

"The post-operative morbidity and mortality of the Fontan operation are still a matter of concern. The purpose of this report is to assess suspected predictors of unstable hemodynamics and death after the Fontan procedure."

catheter. Milrinone was administered for those patients with borderline or increased pulmonary artery pressure. Fontan takedown was considered when patients were not responsive to medical therapy.

Statistical Analysis

Data were presented as mean ± standard deviation for interval and count (percent) for categorical variables. Association between the study outcomes (death and hemodynamic status) and other factors were investigated by student's t test (for interval data) and chi square or Fisher's exact test (for nominal data). P value < 0.05 considered statistically significant. Multivariate analysis was performed using poisson regression models to determine the adjusted associations between favorite outcomes and related factors. We used STATA 8 for Windows (STATA Corporation, Texas, USA) for statistical analysis.

Results

Patients' ages at the time of operation were between 2 and 24 years (mean=8.5±4) with mean weight of 23.5±12 kilograms.

The frequency of female and male sex was 45% and 55% respectively.

Patients consisted of 39 (48.8%) with tricuspid atresia and 41 (51.3%) with single ventricular heart anomaly including those with double inlet, single inlet and common inlet ventricle. No patients with Hypoplastic Left Heart Syndrome were included in our study.

Left ventricular morphology was present in 74 cases (92.5%) with the remainder including those with right ventricular, mixed and undetermined types (7.5%).

Pulmonary stenosis or atresia was present in 74 cases (92.5%).

Patients with severe pulmonary artery branch stenosis did not undergone Fontan operations. Therefore, pulmonary artery branch diameter was acceptable in 60 patients (75%), and relatively small size in the remainder (25%). Nevertheless, all the selected patients had pulmonary artery branch continuity.

Pulmonary artery pressure was less than 15 mm Hg in 72 patients (90%), and in the remainder 10% between 15 and 20 mm Hg.

Mild to moderate atrioventricular valve regurgitation was present in 33 patients (41%), but in the remaining 59%, there was no remarkable regurgitation. Atrioventricular valve repair was performed for 17% of all patients, but no patient with severe atrioventricular valve regurgitation had a Fontan operation.

The position of the great arteries was normal in 44 cases (55%), and transposed to the right or left in the remanding 36 cases (45%). Dextrocardia was present in 7 cases (8.8%).

In 57.5% of all patients, a fenestration was left during the operation. Therefore, it was performed in 61% of atriopulmonary connection, 70% of lateral tunnel and 43% of extra-cardiac conduit.

Previous surgical palliations were performed for 72.5% of all patients, including systemic to pulmonary artery shunt (60%), Glenn shunt (5%) and pulmonary artery banding (7.5%).

June Medical Symposium Focus

Fifth World Congress of Paediatric Cardiology and Cardiac Surgery

June 21-26, 2009
Cairns, Australia
www.PCCS2009.com

PCCS2009 will have a parallel group of eight "Specialty meetings" (Incorporated Satellites) within the framework of the World Congress, followed by three days of academic activity and discussion.

All delegates will be registered for one of the specialty meetings, which will provide a focused start to the *World Congress*.

The Specialty meetings include:

- Pediatric and Interventional Cardiac Symposium (PICS-AICS)
- Surgical (jointly organized with WSPCHS) 2nd Biennial Scientific Meeting of The World Society for Pediatric and Congenital Heart Surgery at the World Congress
- Pediatric Cardiac Intensive Care jointly organized with PCICS. (includes perfusion and anesthesia)
- Adult Congenital Heart Disease jointly organized with International Society for Adult Congenital Heart Disease (ISACHD)
- Pediatric Electrophysiology / Arrhythmia (Mon. and Tue.) jointly organized with PACES and PEDIRHYTHM
- Imaging (including Fetal Echocardiography / MRI and CT)
- Nursing
- Pediatric Cardiomyopathy / Transplant (Monday) Pulmonary Hypertension

Thereafter, the meeting will comprise a series of plenary meetings, each exploring a specific topic, alongside free abstract sessions and educational activities for trainees.

The meeting will conclude on Friday with five "Landmark Lectures" in Pediatric Cardiology, Pediatric Cardiac Surgery, Pediatric Cardiac Intensive Care, Pediatric Cardiac Nursing, and Pediatric Cardiac Pathology

The mean preoperative arterial PO^2 and O^2 saturation were 48 and 77 mm Hg respectively, which increased to 59 and 86 mm Hg after Fontan surgery. The mean PO^2 and O^2 saturation were statistically higher in patients who were provided a lateral tunnel or extra-cardiac conduit (60% and 88 mm Hg respectively) in comparison with atriopulmonary connection (55% and 80 mm Hg) (P=0.03).

Early after Fontan operation 16 patients (20%) needed inotropic agents. Ten patients (12.5%) had unstable hemodynamic in spite of using inotropic agents, but in the remaining 6 cases systemic blood pressure was relatively stable. Eighty percent of those patients with unstable hemodynamic (8 cases) died during the first few hours or days after operation. In comparison, of the 8 patients (33%) who were stable with inotropic agents, 2 cases expired early after surgery.

Mean pump time of patients with unstable hemodynamics (172.2 minutes) was remarkably higher than of the hemodynamically stable cases (122 min). Indicating that hemodynamic instability is statistically related to increased pump time ((P=0.009) Table 1). Likewise, mean pump time of 16 patients who needed inotropic agents (157 min) was statistically higher than of those patients who did not require these drugs (121 min)(P= 0.024).

Other quantitative predictors including age, weight, PO^2 and SaO^2 were assessed for their likelihood association with hemodynamic status, but no statistical relationship was found (Table 1).

Univariate analysis of post-operative hemodynamic status and ventricular morphology showed that non-left ventricular morphology may be a a positive predictor for unstable hemodynamics (P=0.018) (Table 2). Likewise, this relationship

Table 1. Relationship of quantitative variables with death and hemodynamic

Variables	Mean	Hemodynamic		P value	Death		P value
		Stable	Unstable		No	Yes	
Age	8.5±4 y	8.6±3.6	7.7±6	0.5	8.5±3.5	8.5±6	0.9
Wt	23±12 Kg	24±12	20±12	0.3	24±12	23±14	0.8
PO_2	48±9 mm Hg	48±9	48±14	0.9	48±8	50±15	0.3
SaO_2	76±9%	77±9	76±9	0.7	76±8	78±10	0.6
P-time	128±57 min	122±54	172±54	0.009	120±54	173±54	0.002

Table 2. Relationship of cardiac anomaly and outcomes

Predictors		P value	Outcome		P value	Hemodynamic	
			Died	Alive		Unstable	Stable
Yes	TA	0.594	5	34	0.933	5	34
No			7	34		5	36
LV	Vent Morph	0.002	8	66	0.018	7	67
Other			4	2		3	3
Two	AV value	0.196	7	26	0.551	5	28
One			5	42		5	42
Yes	NRGA	0.122	9	35	0.33	7	37
No			3	33		3	33
Yes	Dex	0.956	1	6	0.883	1	6
No			11	62		9	64
Normal	PA branch	0.041	6	54	0.051	5	55
Small			6	14		5	15
No	Regurg	0.719	11	60	0.892	9	62
Yes			1	8		1	8
Yes	PS	0.033	9	65	0.163	8	66
No			3	3		2	4
Normal	M_PAP	0.001	7	65	0.001	5	67
Increased			5	3		5	3

TA=tricuspid atresia, Vent morph=ventricular morphology, AV valve=atrioventricular valve, NRGA=normal related great artery, Dex=dexterocardia, PA branch=pulmonary artery branch, Regurg=atrioventricular regurgitation, PS=pulmonary stenosis, M_PAP=mean pulmonary artery pressure

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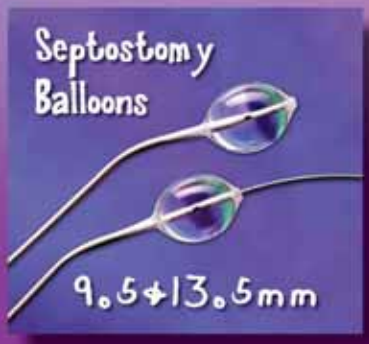

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March Medical Symposium Focus

American College of Cardiology's Congenital Cardiology Solutions (CCS.09)

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At the American Cardiology Scientific Sessions (ACC.09), the focussed CCS. 09 track offers opportunity to learn about congenital heart

Highlighted main symposia topics include:

- Ebstein's Anomaly: Neonate to Adult
- Fontan Late Outcome and Management of the Failing Circulation
- Complex Transposition of the Great Arteries
- Staging the Patient with Hypoplastic Left Heart Syndrome
- Neurodevelopmental Outcomes of Neonates and Infants Undergoing Cardiac Surgery.

Each topic will include the input of cardiologists and surgeons with expertise in each area to discuss management options, imaging, outcomes, and sequelae of the different conditions. The symposium will include adult congenital heart disease for the general cardiologist community.

One oral abstract session will highlight the most pertinent abstracts accepted for CCS.09. Other abstracts will be presented in poster formats. The topics include aortic dilation in pregnant women with Marfan Syndrome, left ventricular dysfunction in tetralogy of Fallot and outcomes of the neoaorta after the arterial switch operation.

Five *Meet the Experts* sessions will provide direct interaction with experts on including advances in fetal diagnosis and management, cyanotic CHD in the adult, and advances in mechanical cardiopulmonary support.

Table 3. Relationship of previous operation and Fontan with outcomes

Predictors		Hemodynamic		P value	Output		P value
		Stable	Unstable		Alive	Dead	
Palliation	yes	48	10	0.088	46	12	0.021
	no	22	0		22	0	
Type of Pre op	shunt	41	7	0.082	40	8	0.04
	Pa band	4	2		3	3	
	Glen	3	1		3	1	
	non	22	0		22	0	
Types of Fontan	AV connection	10	3	0.037	9	4	0.145
	Extra conduit	20	1		20	1	
	Intra cardiac	31	6		30	7	
	Bilateral tunnel	9	0		9	0	

Table 4. Relationship of predictors with outcomes by using multivariate analysis

Variables	Death		Unstable Hemodynamic	
	P value	Coef	P value	Coef
Age	0.55	0.04	0.58	-0.5
PS	0.13	1.2	0.66	0.6
Vent morph	0.049	0.67	0.86	0.1
m-PAP	0.005	2.6	0.001	4.6
Regurgitation	0.5	-0.73	0.16	-2.5
HO palliation	0.99	-17	0.59	-3
Fontan OP	0.035	-1.2	0.006	-1.4
_cons	0.99	13	0.532	2

PS=pulmonary stenosis, OP=operation, HO=history of operation, Coef=coefficient

was present for increased pulmonary artery pressure (P<0.001); whereas, it was borderline for small size pulmonary artery branches (P=0.051) (Table 2).

However, there was no statistical relationship between hemodynamic status and other aspects of cardiac anatomy including: position of great arteries, pulmonary stenosis, cardiac situs and atrioventricular anatomy or regurgitation (Table 2).

Comparing the type of operation with unstable hemodynamics displayed significantly higher occurrence among patients with atriopulmonary connection (P=0.037) (Table 3).

Twenty percent of all patients (16 cases) died after Fontan operation. A review of the time of death statistics revealed that 75% (12 cases) of all mortalities emerged during the first month after operation, whereas just 2 cases (12.5%) died during the following two years after surgery

"In conclusion, shorter pump time and aortic cross-clamp time diminish early post-operative complications and mortality."

indicating that the main cause of death was due to early postoperative complications.

Our review indicated relatively more occurrence of mortality among patients who had undergone atriopulmonary connection (31%) (Table 3). Likewise, of 37 patients with lateral tunnel procedure 7 cases died (19%). In comparison, there was only one death after extra-cardiac conduit operation (4%).

Univariate analysis did not show a relationship between previous history of palliation and unstable hemodynamics (P=0.088), however, previous palliation and death were associated (P=0.04) (Table 3), which could be attributed to pulmonary artery banding.

Pump time had a significant relationship to post-surgical death (P=0.002) (Table 1). Likewise, mean pulmonary artery pressure was statistically related to postoperative death (P<0.001) (Table 2). Other quantitative variables including age, weight, height and preoperative blood O^2 content were not associated with increasing mortality (Table 1).

Considering the kind of heart anatomy indicated that there is no significant statistical relationship between death and tricuspid atresia with transposition of the great arteries, dextrocardia, and atrioventricular anatomy or valve regurgitation. However, this relationship was present between death and ventricular morphology through univariate analysis (P=0.002) (Table 2).

Likewise, the absence of main pulmonary artery stenosis or atresia, was statistically associated with increased mortality (P=0.033) which might be due to increased mean pulmonary artery pressure (Table 2). In comparison, small pulmonary artery branch size was statistically associated with increased mortality (P=0.041) (Table 2).

Poisson regression models were used to determine adjusted association between study outcomes and the predictors which are indicated in Table 4.

Multivariate analysis indicated that increased pulmonary artery pressure had significant association with mortality (P<0.001 with coefficient of 2.6) and hemodynamic instability (P<0.001 with coefficient of 4.6).

According to the adjusted analysis, there was no association between unstable hemodynamics and ventricular morphology (P=0.086); however, a statistical relationship was present between death and ventricular morphology (P=0.049 with coefficient of 0.67) (Table 4).

Other variables which are enrolled in this adjusted analysis including age, main pulmonary artery stenosis, atrioventricular regurgitation and previous history of palliation were not found to be associated with increased mortality or unstable hemodynamic (Table 4).

Discussion

Early after Fontan operation is the crucial period of patients' hemodynamic adaptation with the newly established cardiovascular anatomy. This is demonstrated in our study by occurrence of the majority of mortality during the early post-operative period. Another report showed 22 death (9.8%) early after the Fontan operation.[5]

Although the mean age and weight of patients with unstable hemodynamics were lower, nevertheless, they were not statistically remarkable. This is also supported by another report.[6]

We could not find statistical association between preoperative O^2 saturation or PO^2 and unstable hemodynamic or death (Table 1). Therefore, severity of cyanosis before surgery may not be a risk predictor of the Fontan procedure.

Pump time was an important predictor of hemodynamic disturbances and death. Thus, increasing the pump time was positively related to these undesirable outcomes. Jacobs ML and colleagues[3] noted the definite adverse effect of pump and cross clamp time prolongation on pulmonary, cardiovascular, renal or central nervous systems complications. Therefore, avoidance of aortic cross-clamping and deep hypothermic circulatory arrest is considered important.[3] Adachi I and colleagues[7] believe that off-pump Fontan obviates adverse inflammatory reaction on pulmonary circulation.

Although there are some reports about postoperative effects of fenestration including pleural effusions and hospital stay[6,8], we could not find this association.

Association of left ventricular morphology and increased mortality was indicated by means of multivariate analysis; nevertheless, other reports do not support our result.[6, 9]

Suitable pulmonary resistance either by means of lower pressure or normal branch diameters and distribution is essential for improving postoperative outcome.[2,8] Although, mean pulmonary artery pressure was under 15 mm Hg in the majority of patients and less than 20 mm Hg in the remainder, nevertheless, our study indicated that even a little increase over 15 mm Hg might jeopardize the post-operative patients' status. Hosein RB.M 6

PICS-AICS 09

Pediatric and Adult Interventional Cardiac Symposium

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- **ABSTRACTS** - Interventional abstracts will be presented at the World Congress meeting June 22-26, 2009.

REGISTRATION INFORMATION
This year's PICS~AICS meeting will take place June 21-23, 2009 in Cairns, Australia. PICS~AICS is a sub specialty meeting of the World Congress meeting. For more information go online to: **www.picsymposium.com**

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and colleague reported that increased mean pulmonary artery pressure (>15 mm Hg) was definitely associated with abnormal hemodynamic status and death.[6] Higher mortality in patients with previous pulmonary artery banding might be due to higher mean pulmonary artery pressure before the operation.

Some types of Fontan procedure were associated with increased adverse outcomes, especially when an adjusted multi-variable analysis was made. Thus, higher frequency of mortality and hemodynamic instability were found in patients with atriopulmonary connection followed by lateral tunnel procedure. Its adverse outcomes were essentially due to right atrial dilation and turbulent circulation instead of straight flow in this chamber which would follow by ineffective atrial inflow and output, higher incidence of arrhythmias and also cloth formation.[11] In our study hemodynamic disturbances and mortality were more frequent in those patients with lateral tunnel procedure in comparison to extra-cardiac conduit; however, it was not statistically significant. Nevertheless, this difference may in part be due to more prolonged pump time in the lateral tunnel operation.

According to some reports, the extra-cardiac conduit coupled with minimal use of extracorporeal circulation is associated with favorable intraoperative hemodynamics, low fenestration rate, minimal risk of thrombosis or stenosis, and minimal early and late rhythm disturbance.[12-14]

In conclusion, shorter pump time and aortic cross-clamp time diminish early post-operative complications and mortality. Extra-cardiac conduit Fontans are more effective and safer due to shorter pump and\or cross-clamp times. Lower pulmonary vascular resistance and larger pulmonary arterial diameter support better post-operative hemodynamics. Because the early post-operative period is crucial for adaptations to new hemodynamics, even small disturbances may increase risk.

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Welcome To Holland

This brief essay by Emily Perl Kingsley, was sent to us by the parent of child with Down Syndrome. We would like to share it with our readers, because it poignantly expresses feelings of many parents who have children with chronic cardiac and neonatal health problems.

I am often asked to describe the experience of raising a child with a disability - to try to help people who have not shared that unique experience to understand it, to imagine how it would feel. It's like this.....

When you're going to have a baby, it's like planning a fabulous vacation trip - to Italy. You buy a bunch of guide books and make your wonderful plans. The Coliseum. The Michelangelo. David. The gondolas in Venice. You may learn some handy phrases in Italian. It's all very exciting.

After months of eager anticipation, the day finally arrives. You pack your bags and off you go. Several hours later, the plane lands. The stewardess comes in and says, "Welcome to Holland."

"Holland?!?" you say. "What do you mean Holland?? I signed up for Italy! I'm supposed to be in Italy. All my life I've dreamed of going to Italy."

But there's been a change in the flight plan. They've landed in Holland and there you must stay.

The important thing is that they haven't taken you to a horrible, disgusting, filthy place, full of pestilence, famine and disease. It's just a different place.

So you must go out and buy new guide books. And you must learn a whole new language. And you will meet a whole new group of people you would never have met.

It's just a different place. It's slower-paced than Italy, less flashy than Italy. But after you've been there for a while and you catch your breath, you look around.... and you begin to notice that Holland has windmills....and Holland has tulips. Holland even has Rembrandts.

But everyone you know is busy coming and going from Italy... and they're all bragging about what a wonderful time they had there. And for the rest of your life, you will say "Yes, that's where I was supposed to go. That's what I had planned."

"I am often asked to describe the experience of raising a child with a disability - to try to help people who have not shared that unique experience to understand it, to imagine how it would feel. It's like this....."

And the pain of that will never, ever, ever, ever go away... because the loss of that dream is a very, very significant loss.

But... if you spend your life mourning the fact that you didn't get to Italy, you may never be free to enjoy the very special, the very lovely things ... about Holland.

CCT

Emily Perl Kingsley

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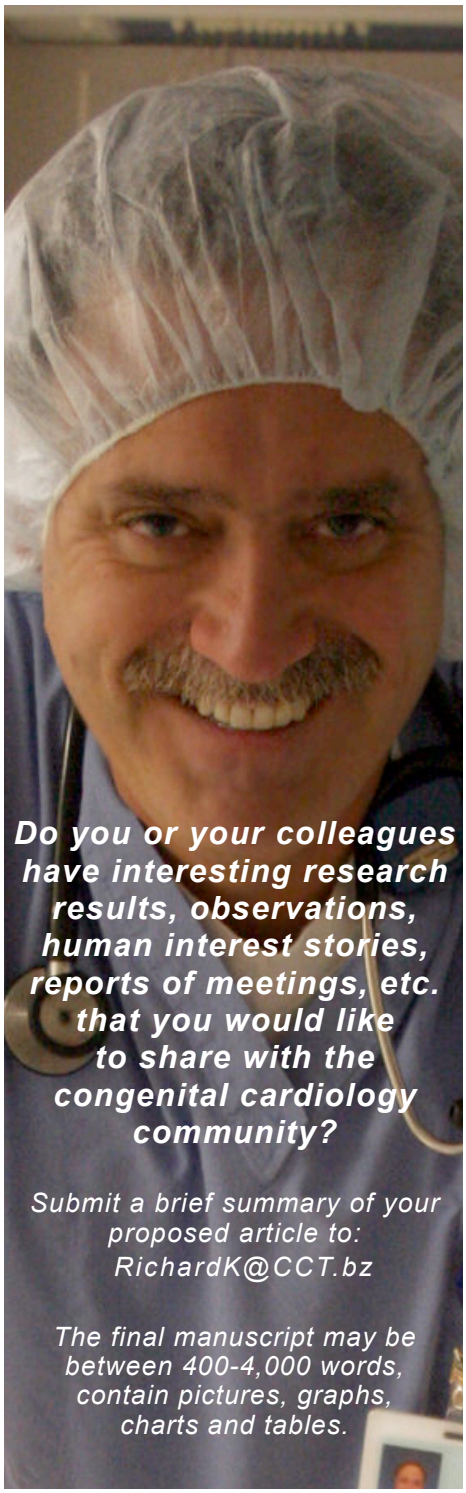

Do you or your colleagues have interesting research results, observations, human interest stories, reports of meetings, etc. that you would like to share with the congenital cardiology community?

Submit a brief summary of your proposed article to: RichardK@CCT.bz

The final manuscript may be between 400-4,000 words, contain pictures, graphs, charts and tables.

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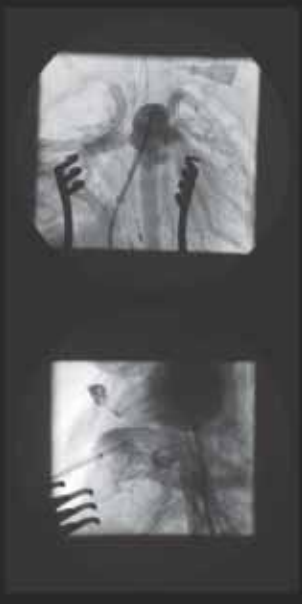
- How to safely manage the physiological stresses of pregnancy
- Best practices for assisting patients and families through critical clinical transitions



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Medical News, Products & Information

First Trial of Gene Therapy for Advanced Heart Failure Shows Promising Results

Newswise — Phase I results of the first clinical trial of gene therapy for patients with advanced heart failure show the approach to be promising, with improvements in several measures of the condition's severity.

In Phase I clinical trials, researchers test a new treatment in a small group of people for the first time to evaluate its safety, determine a safe dosage range, and identify side effects.

Patients enrolled in the multicenter CUPID trial (Calcium Up-Regulation by Percutaneous Administration of Gene Therapy in Cardiac Disease) undergo a minimally invasive cardiac catheterization procedure which introduces a specially engineered gene that stimulates production of an enzyme necessary for the heart to pump more efficiently.

NewYork-Presbyterian Hospital/Columbia University Medical Center was the first to offer the therapy in the New York City area. The Hospital is now recruiting patients for the Phase II CUPID trial to further assess safety and effectiveness in patients with advanced heart failure.

Data from the Phase I trial, which was initiated in May of 2007, were presented at the *American Heart Association (AHA) Scientific Sessions 2008* in New Orleans. Seven of nine patients who were given the drug showed improvements over six months in several areas: symptomatic (five patients), functional (four patients), biomarker (two patients) and left ventricular function/ remodeling (six patients). Two patients with pre-existing antibodies to the viral vector delivery system did not show improvements. Importantly, the approach was shown to have an acceptable safety profile, as determined by an independent safety committee and by the study investigators.

"We are encouraged by these initial findings, which indicate that this therapy has the potential to help patients with advanced heart failure," says Dr. Donna Mancini, the study's principal investigator at NewYork-Presbyterian Hospital/Columbia University Medical Center, where she is Medical Director of Cardiac Transplantation, and is a Professor of Medicine at Columbia University College of Physicians and Surgeons.

The Phase II randomized, double-blind, placebo-controlled clinical trial will compare the therapy at two- or three-dose levels with placebo. CUPID is expected to enroll 46 patients with advanced heart failure at 13 U.S. hospitals.

Gene therapy is a technique for correcting defective genes responsible for disease development by inserting genes into a patient's cells and tissues. In most gene therapy studies, a "normal" gene is inserted into the genome to replace an "abnormal" disease-causing gene. A carrier molecule called a vector must be used to deliver the therapeutic gene to the patient's target cells. Currently, one of the most common vectors is a non-pathogenic virus most people have been exposed to in adolescence that has been genetically altered to carry normal human DNA.

More than five million people in the U.S. have heart failure. Patients with a severe form of the disease have trouble breathing because the heart muscle is not strong enough to pump fluid out of their lungs. Approximately 70% die of the disease within 10 years, and the five-year survival rate is less than 50%. Heart failure is the only cardiovascular disease whose incidence has been increasing rather than decreasing in recent years.

The multicenter national trial is funded and administered by the Celladon Corporation of La Jolla, Calif. (www.celladon.net). The company has reported that the therapy, called MYDICAR®, has been shown to lead to significant improvements in heart function without significant safety concerns in numerous large-animal models of heart failure.

For more information, visit www.nyp.org or www.cumc.columbia.edu

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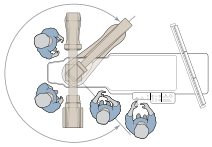

The slender c-arms of our Infinix-i cath lab positioners were built with design input from leading pediatric clinicians, not just engineers in faraway laboratories. Those arms are intricate mechanisms that articulate into optimal positions, yet are simple enough to be driven with one hand. They can be manuevered in the perfect place, out of the way but right where your team needs them for the best possible access to the patient. Discover how Infinix-i can provide the room you need to operate. Get more details at www.medical.toshiba.com.

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