



# Management of Transposition of Great Arteries in Delayed Presenters

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***4<sup>th</sup> Annual Scientific Meeting, Seoul Dragon City, May31st to June 1<sup>st</sup> 2024***



# Disclosures

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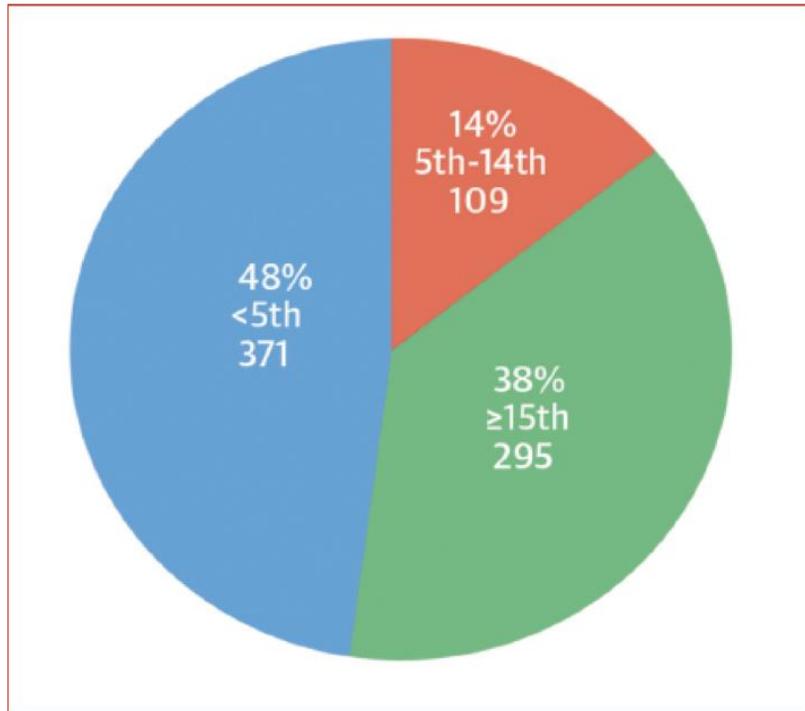
- None

# Transposition of the Great Arteries in the Developing World

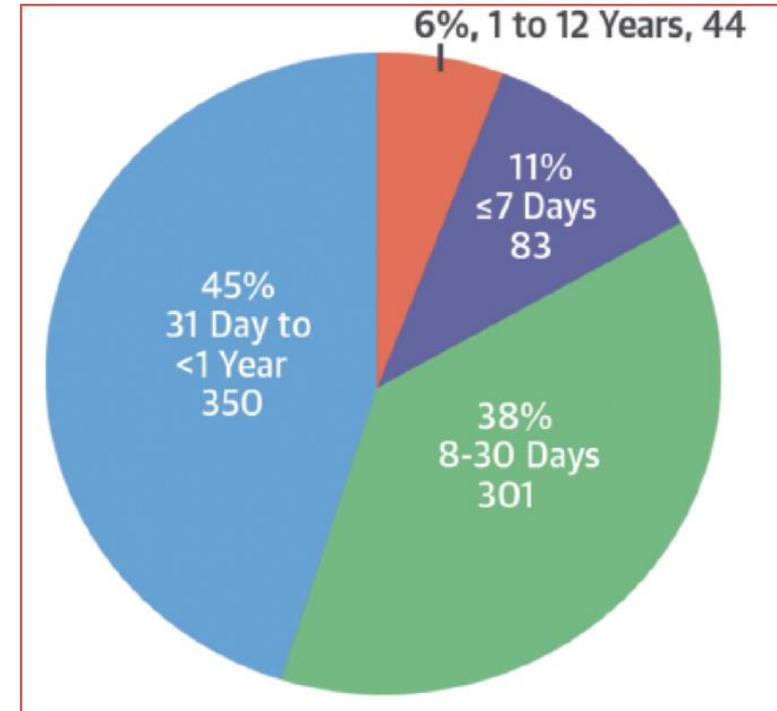
## Surgery and Outcomes

David N. Schidlow, MD, MMus,<sup>a</sup> Kathy J. Jenkins, MD, MPH,<sup>b</sup> Kimberlee Gauvreau, ScD,<sup>b</sup>  
Ulisses A. Croti, MD, PhD,<sup>c</sup> Do Thi Cam Giang, MD,<sup>d</sup> Rama K. Konda, DCH,<sup>e</sup> William M. Novick, MD, MS,<sup>f</sup>  
Nestor F. Sandoval, MD,<sup>g</sup> Aldo Castañeda, MD, PhD<sup>h</sup>

### Weight /BMI for Age Percentile



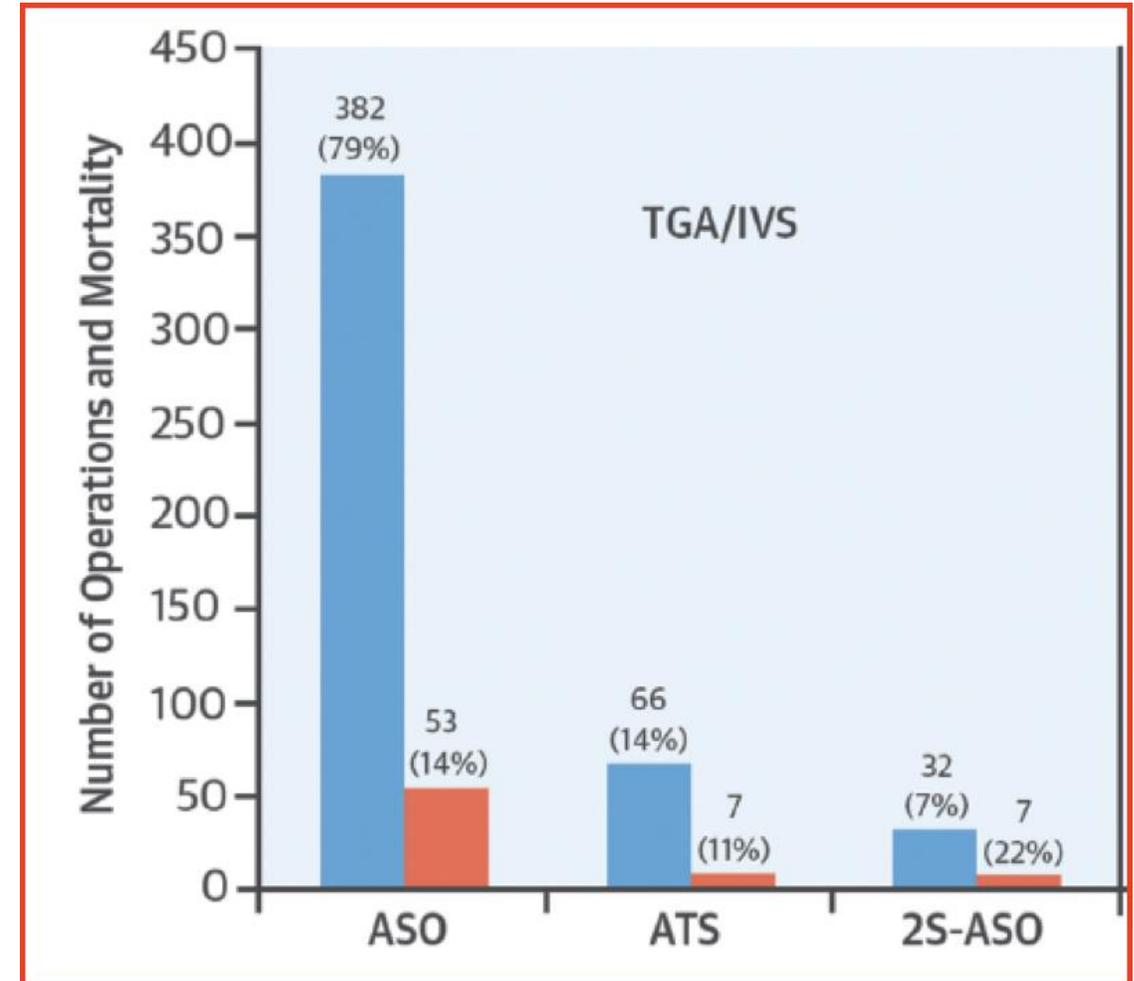
### Age at Surgery



## Multivariate Analysis with Mortality

	OR	95% CI	p Value
<b>Patient characteristics</b>			
WHO weight/BMI-for-age percentile			
<5th	2.23	1.48-3.33	<0.001
5th-15th	1.66	0.91-3.04	0.10
≥15th	1.00	—	—
Male	1.36	1.07-1.75	0.01
<b>Adding annual TGA volume</b>			
WHO weight/BMI-for-age percentile			
<5th	1.98	1.30-3.02	0.002
5th-15th	1.60	0.91-2.82	0.10
≥15th	1.00	—	—
Male	1.45	1.10-1.90	0.008
Average annual volume of TGA repair			
<10	4.71	2.10-10.5	<0.001
10-19	2.41	0.91-6.41	0.08
≥20	1.00	—	—

## In Hospital Mortality



# Reasons for Delayed Presentation

- ✓ Major Medical Illness

**Later Age at Presentation rather than  
Intentional delay in surgical repair**

- ✓ Logistics

- ✓ Pre-op resuscitation

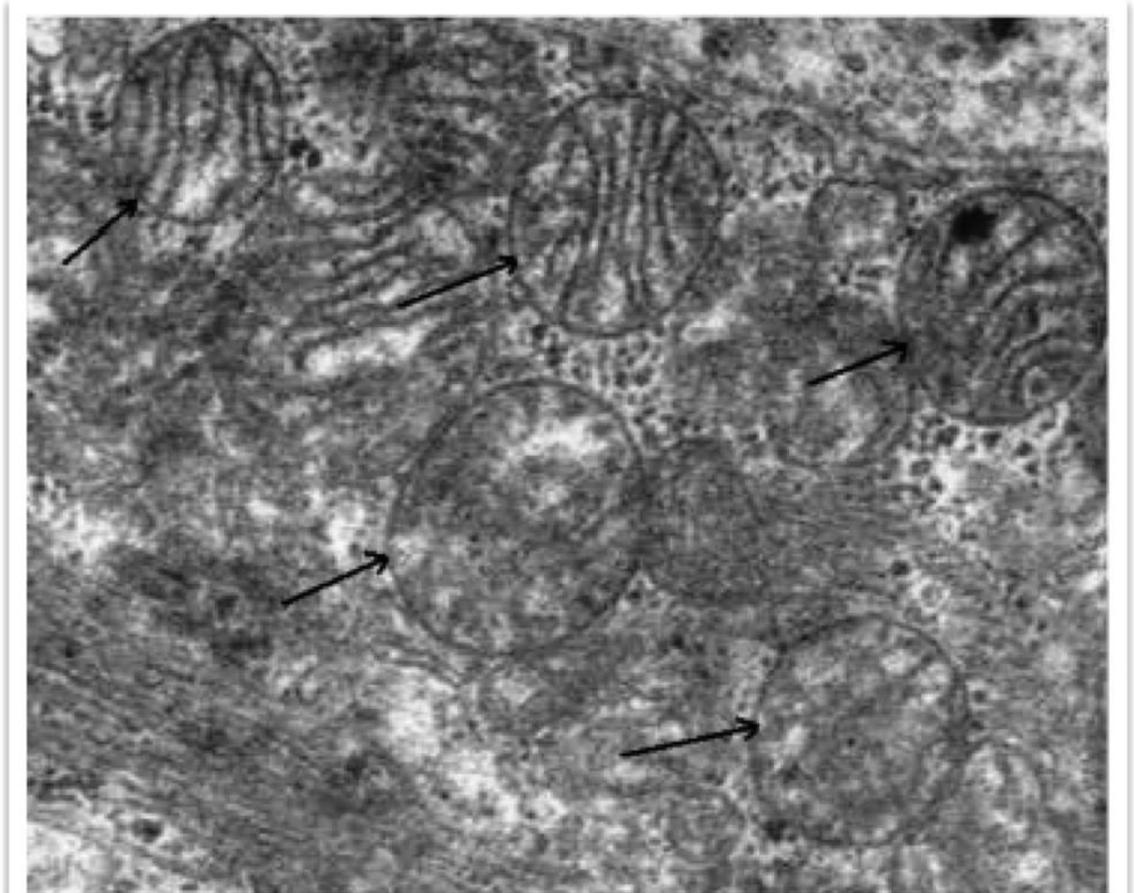
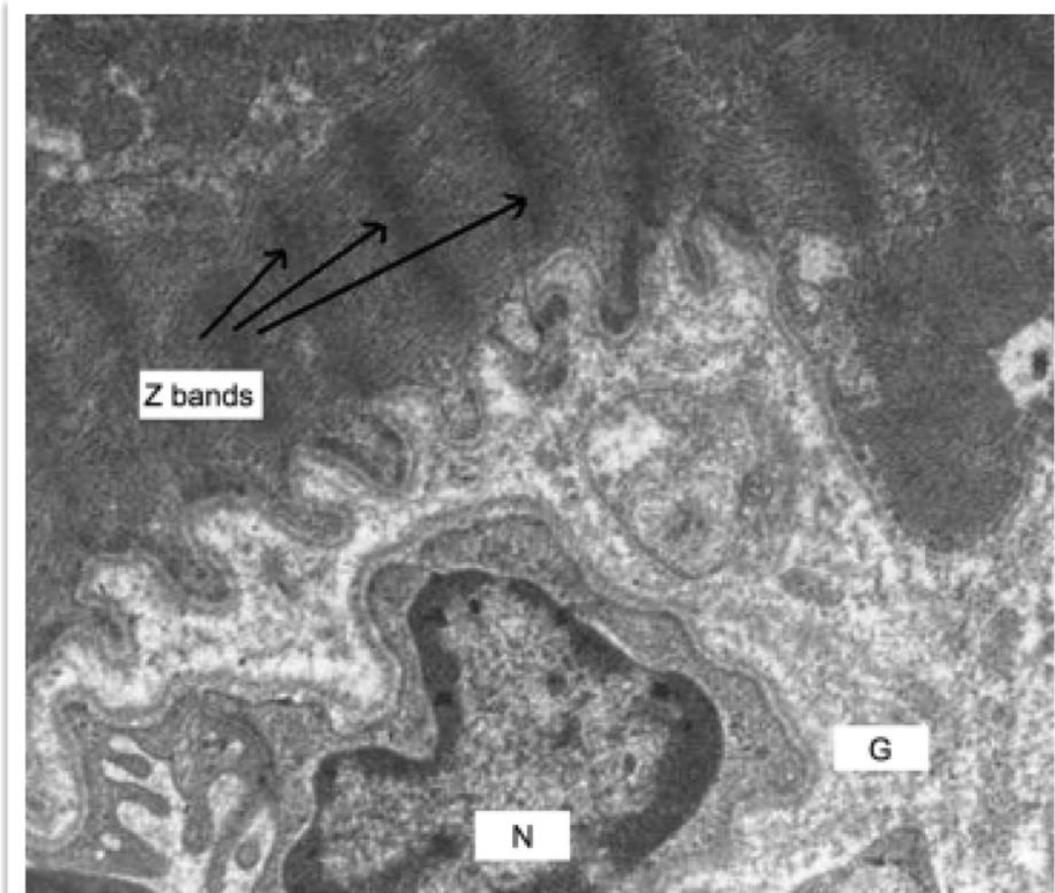
# Considerations in Late Presenters

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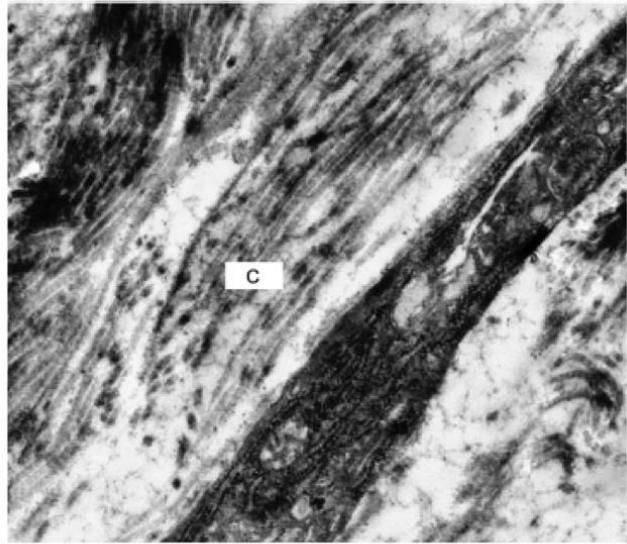
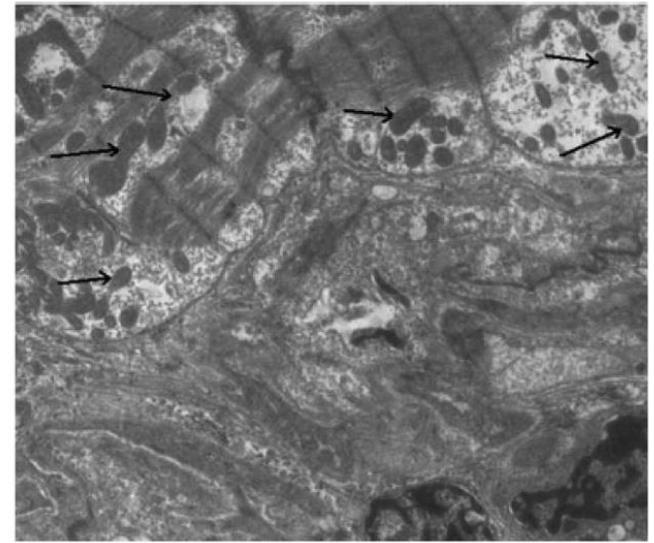
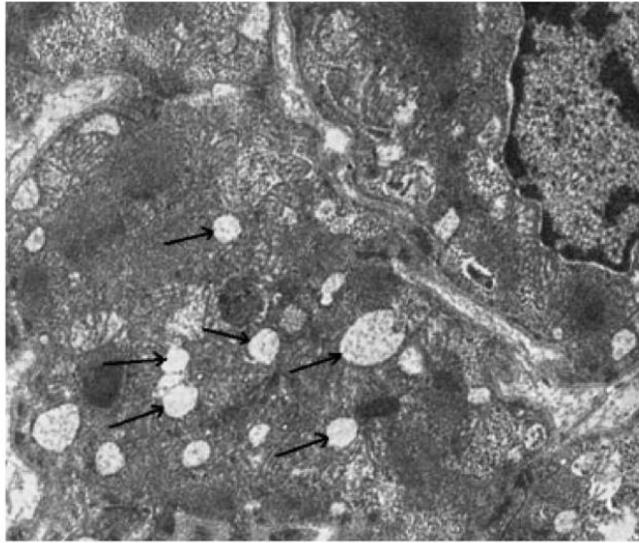
- ✓ Adequacy of left ventricular myocardial mass
- ✓ Assessment of left ventricular preparedness
- ✓ Training for left ventricle for delayed arterial switch
- ✓ Staged Arterial Switch
- ✓ Delayed Primary arterial switch

# Electron Microscopy of LV regression

## Prepared Left Ventricle



# Regressed Left Ventricle



# **Factors Preventing LV regression**

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- ✓ **Isolated Pulmonary outflow obstruction**
- ✓ **Moderately restrictive Atrial Septal Defect**
- ✓ **Sizeable Patent Ductus Arteriosus**
- ✓ **Generally pre-determined factors**

# Surgical management and indication of left ventricular retraining in arterial switch for transposition of the great arteries with intact ventricular septum<sup>☆</sup>

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Received 21 March 2001; received in revised form 27 June 2001; accepted 5 July 2001

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## Abstract

**Objective:** Arterial switch is the operation of reference for the surgical treatment of transposition of the great arteries. In cases of late referral, perinatal complications or early left ventricular (LV) dysfunction, the one stage arterial switch is contra indicated. Anatomical repair remains possible in these patients following a LV retraining. **Methods:** From January 1992 to January 2000, a LV retraining was attempted in 22 patients with transposition of the great arteries with intact ventricular septum (TGA IVS), whereas 470 direct arterial switch and 2 Senning were performed. Indication for LV retraining was based on a combination of factors including: an age older than 3 weeks, a 'banana shape' aspect of the inter-ventricular septum and mainly a LV mass  $<35\text{G/m}^2$ . **Results:** The mean age at LV retraining was 3.2 months ranging from 9 days to 8 months. Usually conducted by sternotomy, it associated a loose PA banding with a LV/RV at 65% with a systemic-pulmonary shunt. The first stage was associated with frequent LV dysfunction and the LV retraining was discontinued in two patients in favor of one Senning and one early switch followed by ECMO. One patient died at first stage from a mediastinitis. Nineteen patients underwent a second stage arterial switch that was performed when the LV mass had reached  $50\text{G/m}^2$  after a mean delay of 10 days, ranging from 5 days to 6 weeks. After a mean follow up of 25 months, there was one non-cardiac late death. The 17 patients followed and leaving with an arterial switch are in NYHA class I, with a mean LV shortening fraction of 39%. **Conclusions:** Arterial switch following LV retraining in TGA IVS is a satisfactory option. The inferior limit of  $35\text{G/m}^2$  adopted, to indicate LV retraining, seems a safe landmark. The quality of the myocardium generated and the respective roles played by the LV afterload, LV wall shear stress, LV inflow and outflow to induce the LV remodeling remain under debate. © 2001 Elsevier Science B.V. All rights reserved.

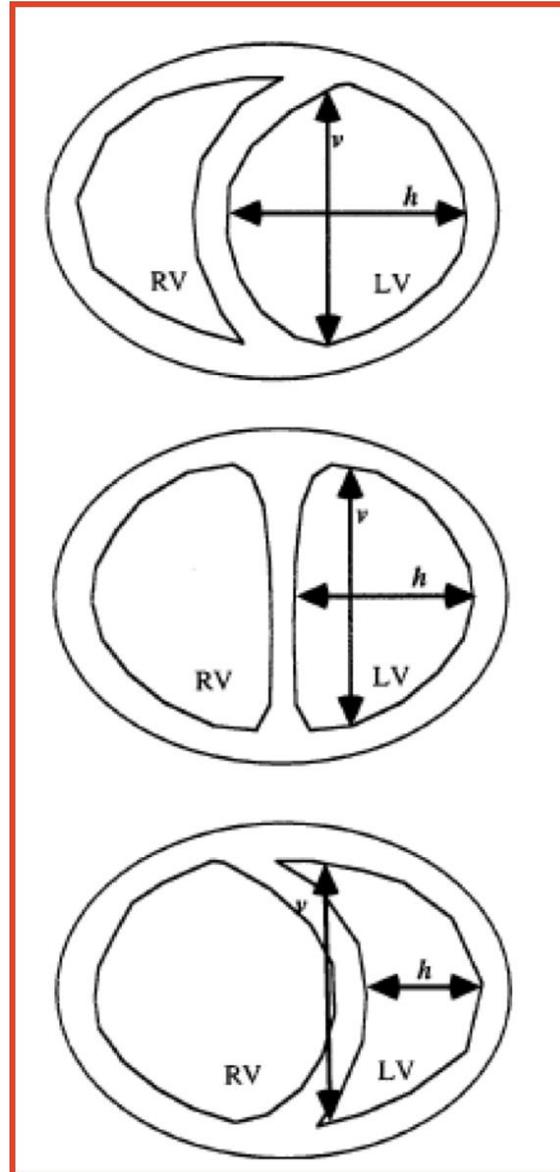
# Indexed Left Ventricular Mass

LV mass (ASE) =  $1.04 * (LVEDd + LVPWd + IVSd)^3 - LVEDd^3$   
Indexed LV mass (G/m<sup>2</sup> =  $0.8 * (LV \text{ Mass}) \text{ to } 0.6 \text{ ] / BSA}$

Indexed LV mass - 35G/m<sup>2</sup> as inferior limit

- ✓ Age > 3 weeks
- ✓ Interventricular septum shape
- ✓ Presence of ASD, PDA and LVOT obstruction

# Patterns of LV Geometry



Favorable (Type I)

Acceptable (Type II)

Unfavorable (Type III)

# Late Primary Arterial Switch

# Extending the Boundaries of the Primary Arterial Switch Operation in Patients With Transposition of the Great Arteries and Intact Ventricular Septum

Nicholas Kang, FRACS; Marc R. de Leval, MD, FRCS; Martin Elliott, MD, FRCS;  
Victor Tsang, MS, MSc, FRCS; Ergin Kocoyildirim, MD; Igor Sehic, MD;  
John Foran, FRCP; Ian Sullivan, FRACP

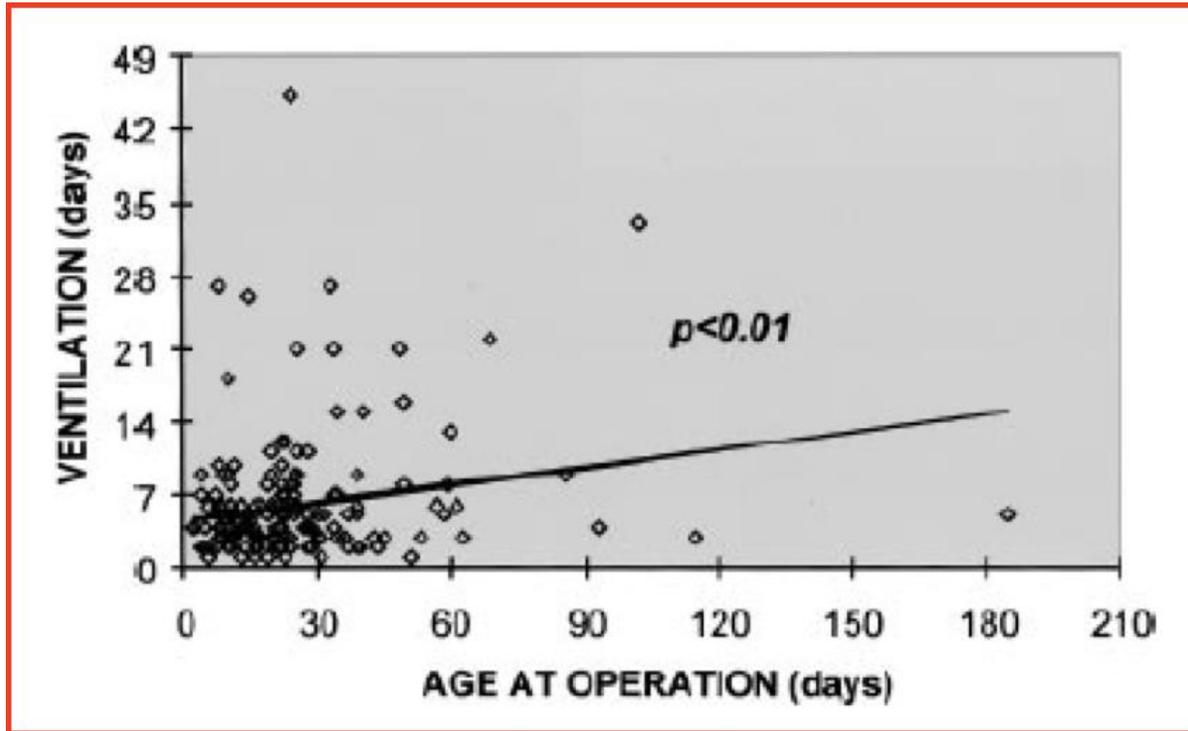
**Background**—We have previously suggested that the primary arterial switch operation is a feasible strategy for patients with transposition of the great arteries and intact ventricular septum (TGA-IVS) up to age 2 months. This study reports our current results with this approach and examines whether this policy could be extended beyond age 2 months.

**Methods and Results**—380 patients who underwent arterial switch for TGA-IVS were reviewed. 275 patients were younger than 3 weeks at the time of surgery (early switch group); 105 patients were 3 weeks or older (range, 21 to 185 days) (late switch group). There was no difference in outcome in terms of in-hospital mortality (5.5% versus 3.8%) or need for mechanical circulatory support (3.6% versus 5.7%) between early and late switch groups. However, duration of postoperative ventilation (4.9 versus 7.1 days,  $P=0.012$ ) and length of postoperative stay (12.5 versus 18.9 days,  $P<0.001$ ) were significantly prolonged in the late switch group. Primary left ventricular failure resulting in death occurred in 2 patients in the late switch group, with no deaths in 9 patients aged 2 to 6 months.

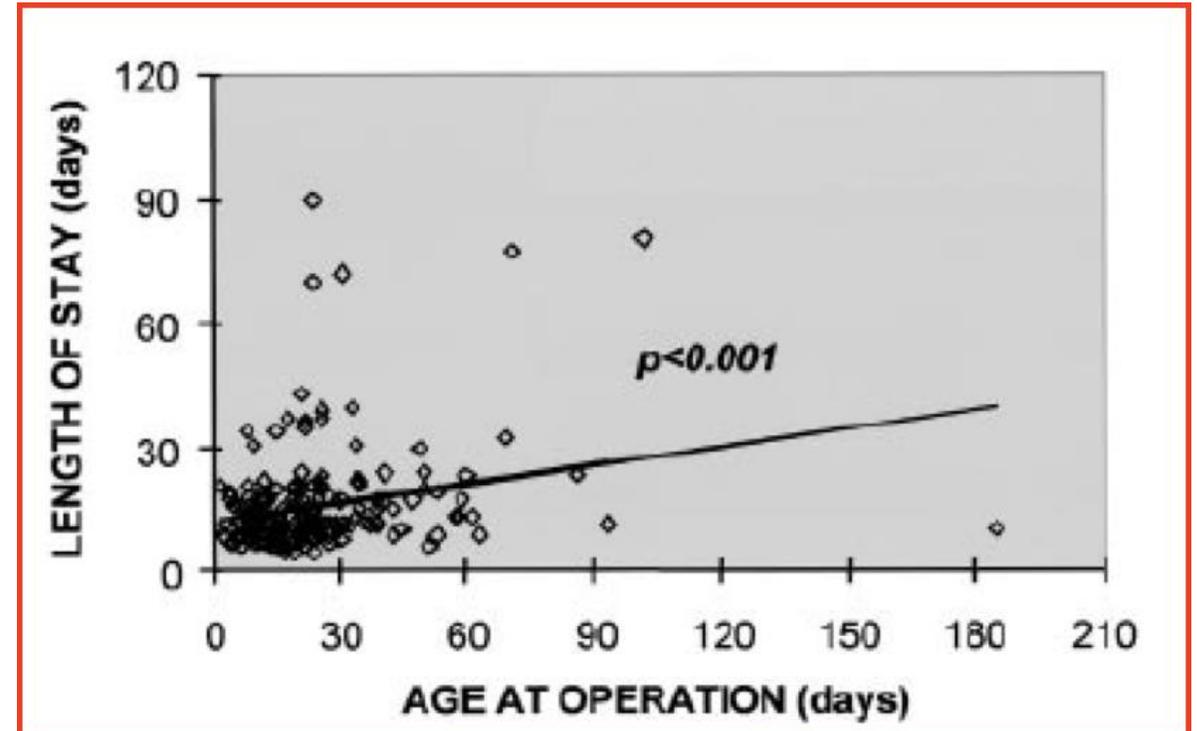
**Conclusions**—This experience confirms that in TGA-IVS, the left ventricle maintains the potential for systemic work well beyond the first month of life. Consequently, neonates at high risk or late referrals can benefit from delayed arterial switch, even beyond age 2 months. However, the need for mechanical support in some of the older patients may limit the widespread adoption of such a strategy. (*Circulation*. 2004;110[suppl II]:II-123–II-127.)

**Key Words:** transposition of great vessels ■ heart defects ■ congenital ■ surgery

## Duration of Post-operative ventilation



## Post-operative length of stay



	Early Switch Group (n=101), d	Late Switch Group (n=101), d	P
Duration of ventilation	4.9±4.3	7.1±7.1	0.012
Length of stay	12.5±6.2	18.9±15.9	0.0003

	Early Switch Group (n=275), %	Late Switch Group (n=105), %	P
Mortality	5.5	3.8	0.51
ECMO (all cases)	3.6	5.7	0.37
ECMO (survived)	2.5	4.8	0.27

# **Training of the left Ventricle**

# Types of Left Ventricular Training

## Hypoxic (Pre-ASO)

- ✓ PA Band with Shunt
- ✓ PA Band with PDA patency
- ✓ Induced Patency of PDA alone

## Normoxic (Post-ASO)

- ✓ Pharmacological Management
- ✓ Mechanical Circulatory Support
- ✓ ECLS
- ✓ VAD

# Primary arterial switch operation in children presenting late with d-transposition of great arteries and intact ventricular septum. When is it too late for a primary arterial switch operation?

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Received 29 November 2009; received in revised form 10 March 2010; accepted 21 March 2010

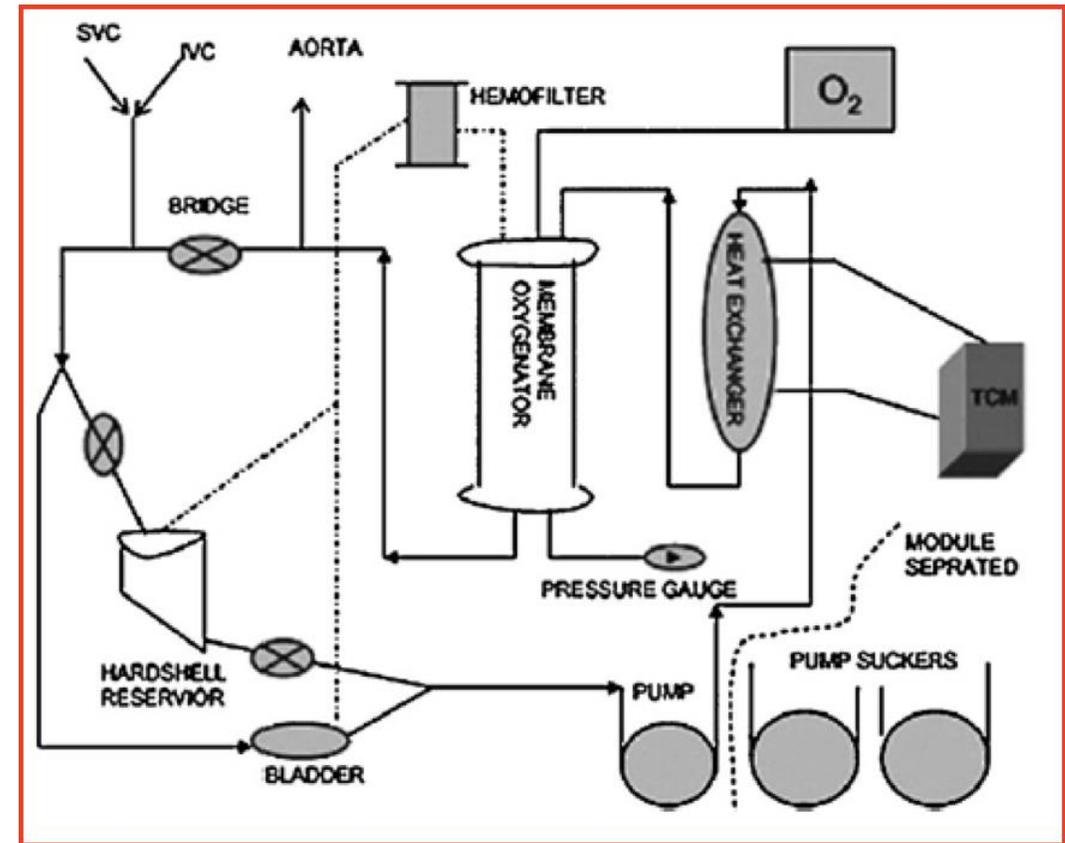
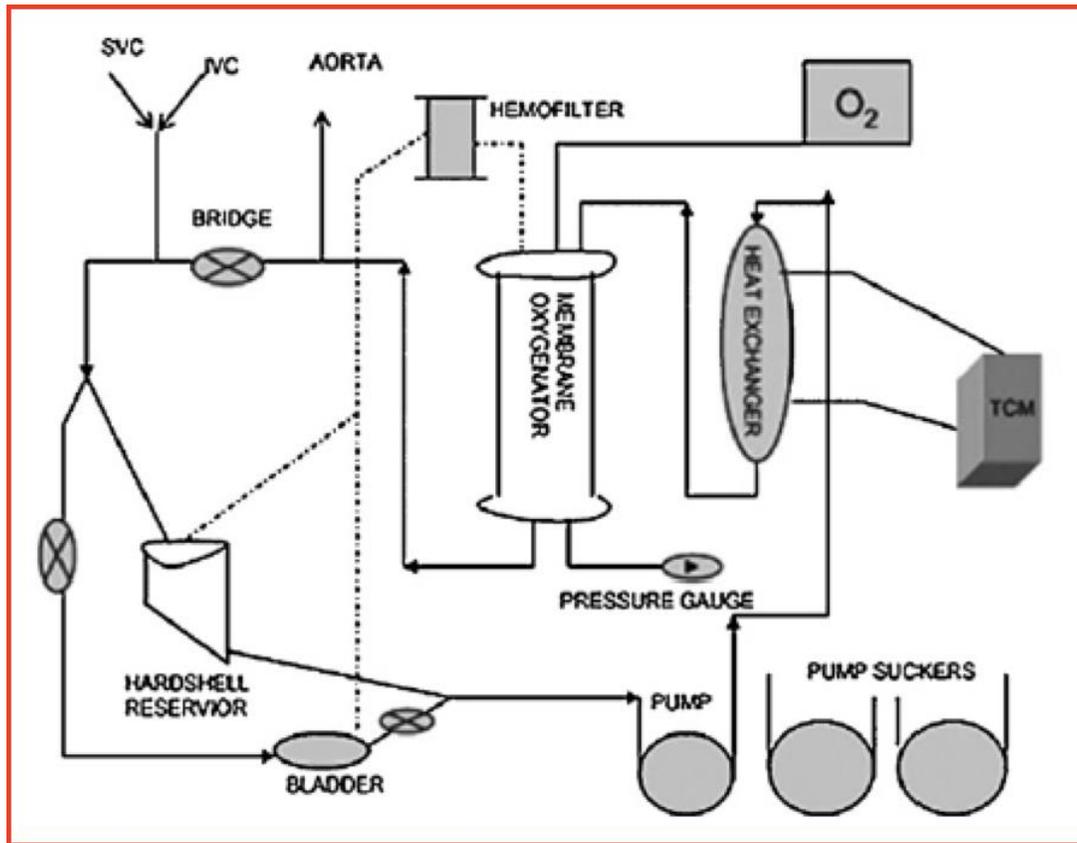
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## Abstract

**Objective:** The surgical management of infants older than 2 weeks with d-transposition of great arteries and intact ventricular septum (IVS) is a matter of debate. Some studies have presented good results of primary arterial switch operation (ASO) in these children. The aim of this study was to assess the surgical outcome of the primary ASO in children with d-transposition of great arteries and IVS presenting beyond 6 weeks of age. **Methods:** The clinical records of the children (more than 6 weeks age) with d-transposition of great arteries and IVS, who underwent primary ASO at our institute between January 2003 and June 2009 were reviewed. Left ventricular geometry and interventricular septal motion on the transthoracic cross-sectional echocardiogram were taken to assess the left ventricle preparedness. **Results:** Fifty-five children (age ranging from 42 days to 9 years) with d-transposition of great arteries and IVS underwent primary ASO. The mean cardiopulmonary bypass time was  $94.7 \pm 21.3$  min, while mean aortic cross-clamp time was  $53.2 \pm 8.1$  min. Seven (13%) of these children died during their hospital stay. The children who had severely regressed left ventricle (banana-shaped left ventricular geometry) were operated with integrated extra corporeal membrane oxygenation—cardiopulmonary bypass (ECMO—CPB) circuit for left ventricular re-training. The children with regressed left ventricle required longer ventilatory time and inotropic support. Recovery of left ventricular geometry has taken 1–6 months depending on age at surgery. **Conclusions:** The children older than 6 weeks with d-transposition of great arteries and IVS can benefit from primary ASO with acceptable results. However, the need for mechanical support in some of the older patients may limit the widespread adoption of such a strategy.

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# Integrated ECMO circuit



## **Use of Left Ventricular Assist Device After Arterial Switch Operation in Late Presenting D-Transposition of Great Arteries—A Technique for Retraining the Regressed Left Ventricle**

**Dhananjay P. Malankar, MCh<sup>1</sup>, Sachin Patil, MD<sup>2</sup>, Shivaji Mali, DNB<sup>2</sup>, Shyam Dhake, DNB<sup>2</sup>, Bharat Soni, MCh<sup>1</sup>, Dinesh Kandavel, PA<sup>1</sup>, Amit Mhatre, MD<sup>2</sup>, Dilip Bind, MD<sup>2</sup>, and Swati Garekar, ABPC<sup>3</sup>**

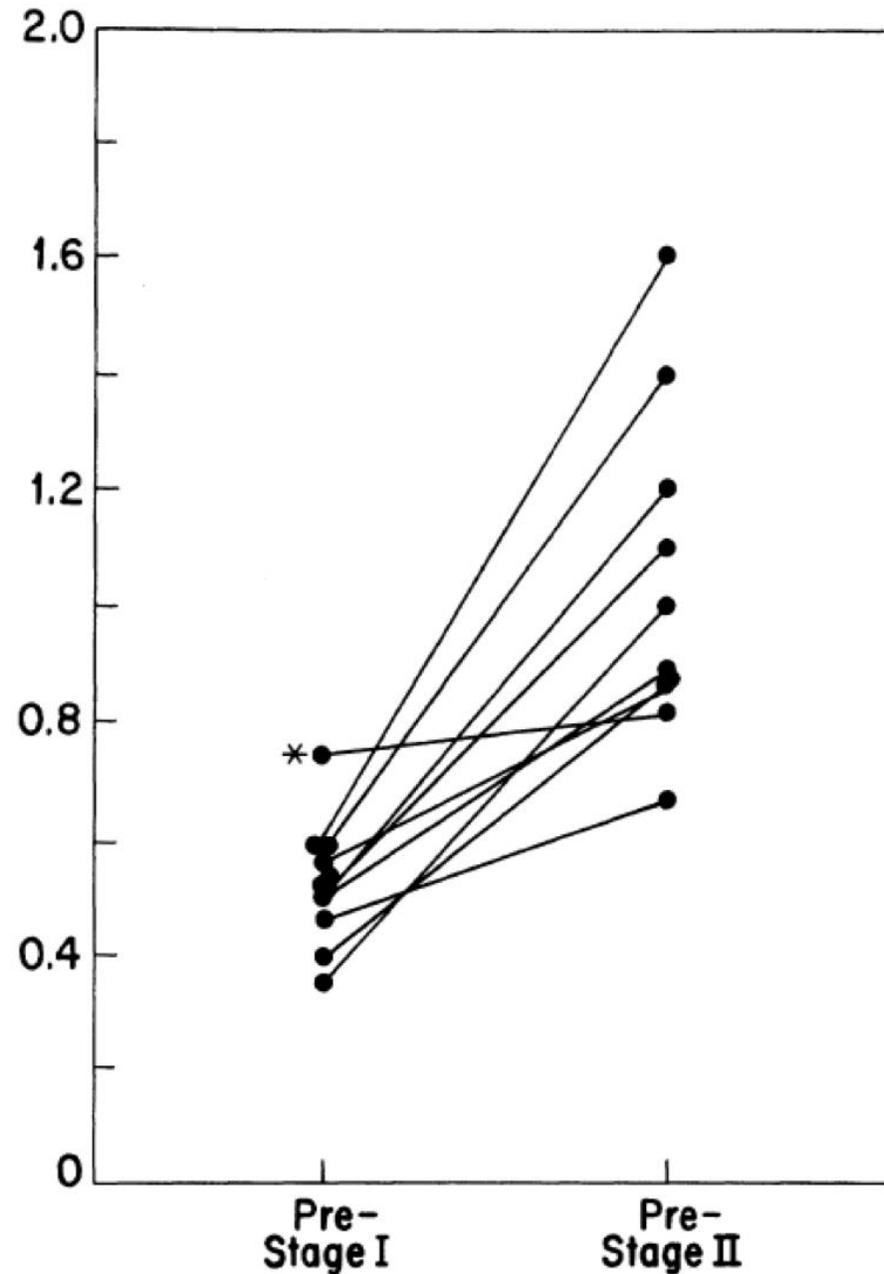
- Lower ACT
- Maximum Left ventricular Decompression
- Technically easier
- Less intensive post operative management

# Pressure Loading LV training

# **Rapid, Two-Stage Arterial Switch for Transposition of the Great Arteries and Intact Ventricular Septum Beyond the Neonatal Period**

Richard A. Jonas, MD, Therese M. Giglia, MD, Stephen P. Sanders, MD,  
Gil Wernovsky, MD, Bernardo Nadal-Ginard, MD,  
John E. Mayer Jr., MD, and Aldo R. Castaneda, MD

**Optimal management of dextrotransposition of the great arteries with intact ventricular septum is currently an arterial switch procedure performed in the first 2 weeks of life. However, a subgroup of patients presents for surgery beyond this time for reasons of sickness, size, or late referral. Experience with 11 such patients (mean age at first-stage procedure, 4.5 months) has revealed that the left ventricle can be prepared by a surprisingly short interval period (median, 9 days) between a first-stage preparatory operation (pulmonary artery band with or without a shunt) and a subsequent second-stage arterial switch procedure. Serial two-dimensional echocardiography showed that left ventricular mass increased by a mean of 85% during this short interval. Mean left ventricular-right ventricular-pressure ratio as measured by cardiac catheterization increased from  $0.5 \pm 0.08$  a median of 7 days before the first stage to  $1.04 \pm 0.29$  a median of 7 days after the first stage. One patient underwent a Senning procedure because of an intramural left coronary artery. The other 10 patients underwent an arterial switch, with no early deaths. Median hospitalization after the arterial switch was 8 days. There has been one late death at 5 months. No patient has been detected to have abnormal ventricular function, although trivial to mild aortic regurgitation has been commonly observed with color flow mapping. These results have encouraged us to offer a two-stage arterial switch procedure to appropriate infants with an interval period of approximately 1 week. Both stages can be performed at one hospitalization, with important psychosocial, logistic, and financial advantages. (*Circulation* 1989;80(suppl I):I-203-I-208)**



### Parameters

LVEF = 42+/-9% - 78+/-9%

LV vol = 52+/-31.7 - 59+/-20.7 ml/m<sup>2</sup>

LV mass = 34.9+/-12.7 - 62.3+/-12.3 g/m<sup>2</sup>

Mass to Vol. ratio = 0.74+/-0.18 - 1.01+/-0.18

# Indications for Two Stage Arterial Switch Operation

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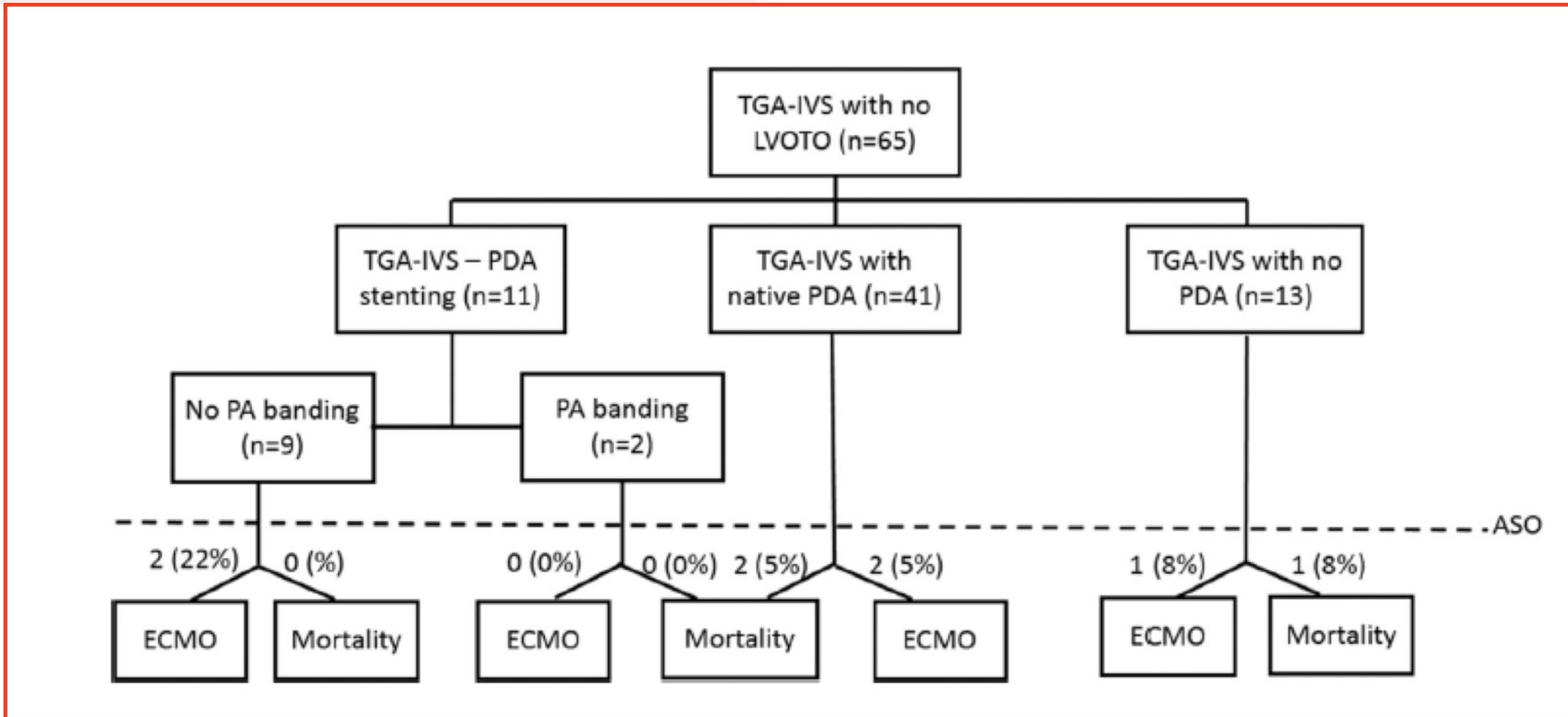
- ✓ Indexed left ventricular mass  $<35\text{g}/\text{m}^2$
- ✓ Age well above 3 weeks
- ✓ Banana-like left ventricular shape
- ✓ Absence of a patent arterial duct or LVOT obstruction

# **Volume Loading for LV training**

# Ductal Stenting to Retrain the Involved Left Ventricle in D-Transposition of the Great Arteries

Ming Chern Leong, MRCPCH, Abdalrahman Ali Ahmed Alhassan, MBBS, Sivakumar Sivalingam, FRCS, and Mazeni Alwi, MRCP

Paediatric & Congenital Heart Centre, Institut Jantung Negara (National Heart Institute), Kuala Lumpur, Malaysia



# Variables for Successful and Failed LV training

Baseline Variables	Successful Retraining	Failed Retraining	<i>p</i> Value
Age at stenting, days			0.545
<42	3 (50)	3 (50)	
≥42	4 (80)	1 (20)	
Weight at stenting, kg			1.000
<3.3kg	3 (75.0)	1 (25.0)	
≥3.3kg	4 (57.1)	3 (42.9)	
Duration of stenting, days			0.242
<163	5 (83.3)	1 (16.7)	
≥163	2 (40.0)	3 (60.0)	
SpO <sub>2</sub> %			0.675
<75	2 (50)	2 (50)	
≥75	5 (71.4)	2 (28.6)	
ASD/IAS			0.018
<0.38	6 (100)	0 (0)	
≥0.38	1 (20)	4 (80)	

- ✓ Easier management in post procedure
- ✓ Minimal inotrope requirement
- ✓ Short ICU and hospital stay
- ✓ Feasible to recanalize the closed ductus

# Ductal Stenting to Permit Delayed Arterial Switch Operation in a Separated Conjoined Twin



Maruti Haranal, MCh, Ming Chern Leong, MRCPCH,  
Siva Rao Muniandy, MRCPCH,  
Khairul Faizah Khalid, MMed, and  
Sivakumar Sivalingam, FRCS(CTh)

# Case Summary

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**Thoraco-omphalophagus Twins**

**Full term via. Caesarean section**

**Twin1 - Normal Heart**

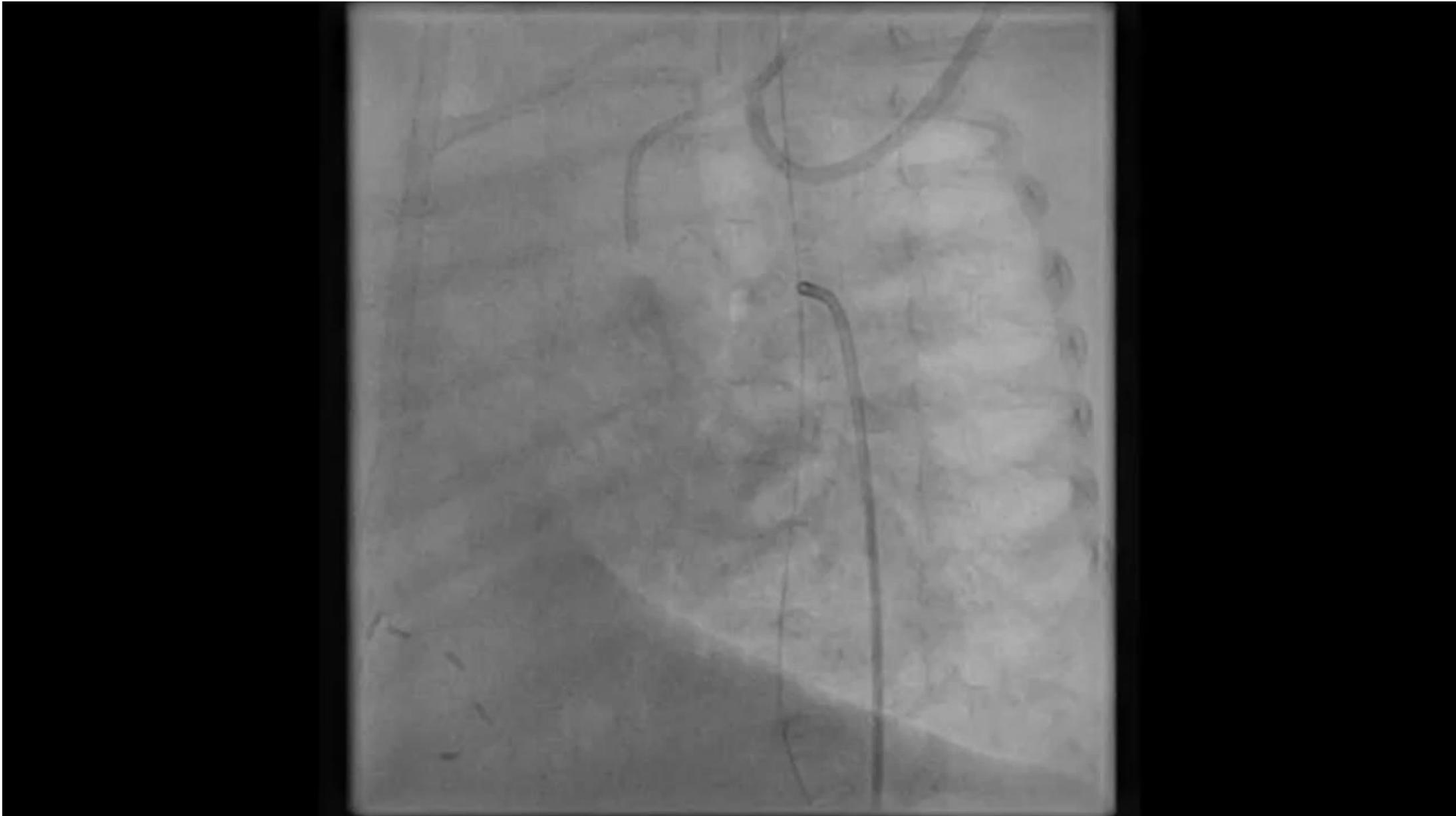
**Twin 2 - Dextrocardia, Transposition  
and intact ventricular septum**

**Rapid Desaturation in Twin 2**

**Commenced of Prostaglandin infusion**

**Teleconferencing with IJN and advised for  
Balloon Atrial Septostomy**





- ✓ 3 months of age
- ✓ High CRP - 45mg/L
- ✓ Tracheal culture - *Acinetobacter* sp.
- ✓ Pulmonary hypertension
- ✓ generalised edema



FR 61Hz  
9.0cm

2D  
81%  
C 44  
P Low  
HGen

M3



JPEG

125 bpm



PR 01HZ  
7.0cm

2D  
73%  
C 50  
P Low  
HGen



100

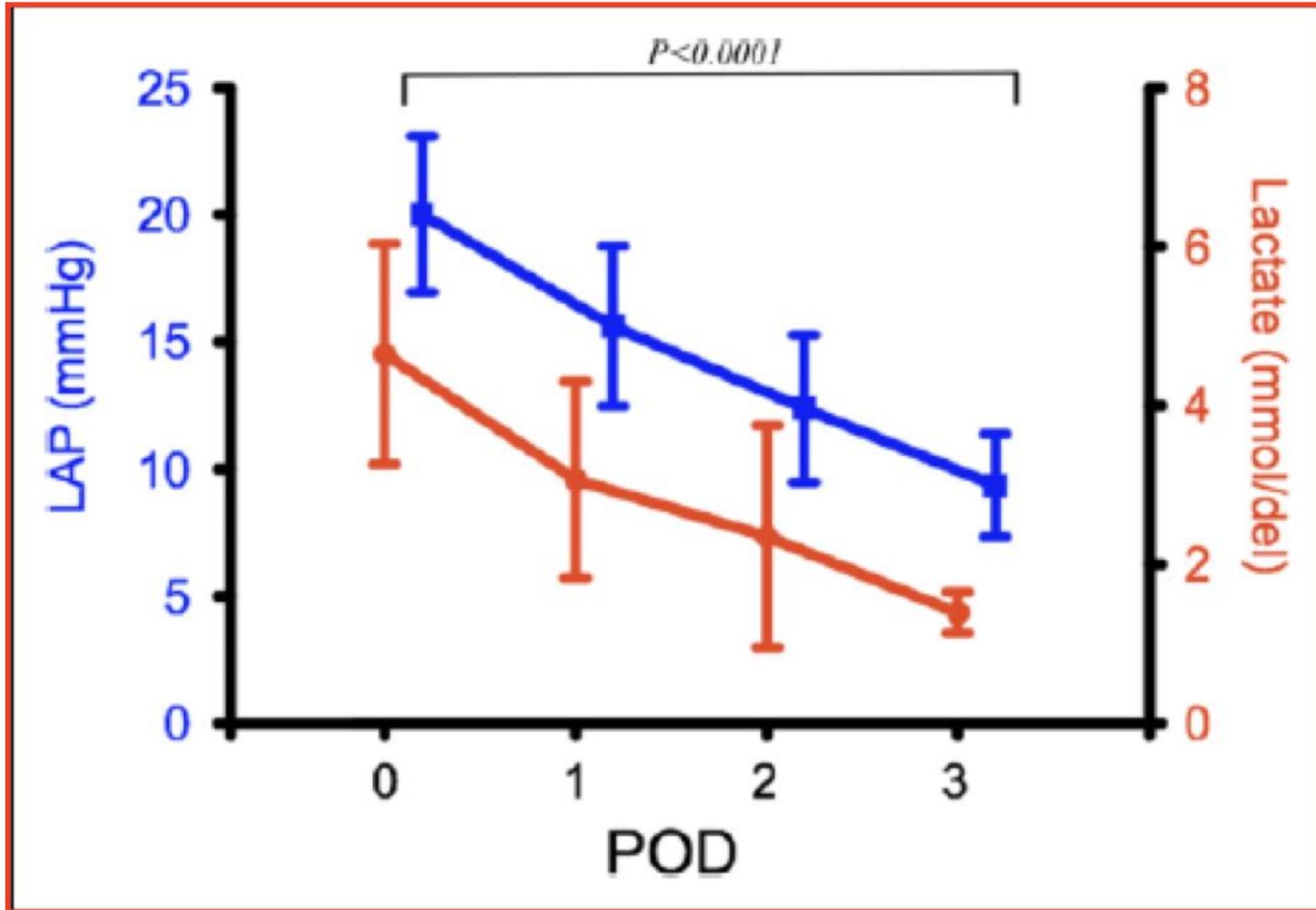
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\*\*\* bpm

# **Post operative Parameters**

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- ✓ **Accept low Blood pressure**
- ✓ **Monitor cerebral and somatic oximetry**
- ✓ **Monitor LA pressure**
- ✓ **Optimal inodilator agent – Milrinone**
- ✓ **Prophylactic peritoneal dialysis**
- ✓ **Delayed Sternal Closure.**



# **Take Home Message**

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- ✓ **Delayed Arterial Switch is feasible in late presenters with TGA,IVS**
- ✓ **Volume Loading strategy is preferable for training the Left ventricle**
- ✓ **Post operative strategies need to be tailored for successful outcomes**
- ✓ **Need to mechanical support should be given consideration in selective cases**
- ✓ **Long term outcomes with objective measures need to be considered**



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