Evaluation of early repair of Complete Atrio-ventricular Canal Malformations

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Abstract

Background & Objective: Since the first documented description of CAVCD repair by Lillehei and colleagues in1954, various techniques have been proposed for its repair. However, the ideal timing for the repair remains controversial. Many authors now advocate primary repair at the age of six months or earlier before the onset of irreversible pulmonary hypertension especially in the presence of Down's syndrome. The aim of this work is to evaluate the results of early total repair of CAVCD before six months of age with the hypothesis that, with modern techniques, the current risks of CAVSD repair in children younger than 6 months and those older than 6 months are equal.

Patients & Methods: 20 patients (11 females 9 males) below 6 months of age with isolated CAVC *(study group*) who underwent primary total repair were included. Another group of 20 patients (12 females & 8 males) with CAVC who underwent repair above the age of 6 months were also selected for comparison (*control group*). The mean age in each group was 5.3$\pm$0.49 & 12$\pm$ 8.5months respectively while the mean body weight was 4.7 $\pm$ 0.45 & 9$\pm$ 2.5 Kg respectively. Down syndrome was present in 9 patients of the study group (45%) and in 10 patients (50%) of the control group.

Results: There was no statistical difference in intra-operative and postoperative data between both groups, apart from: the duration of mechanical ventilation which was longer in the control group (48 $\pm$ 20 hours) than in the study group (36 $\pm$ 15 hours) and the incidence of recurrent postoperative pulmonary hypertensive crises which was higher in the control group (8 patients) than in the study group (4 patients).On the other hand the durations of ICU & hospital stay were higher in the study group (5$\pm$1 &10$\pm$ 1.5 days respectively) than in the control group (4$\pm$0.88 &9$\pm$ 1.27days respectively. .

Conclusion: We concluded that with modern techniques, the current risk factors for CAVSD repair in patients younger than 6 months and in those older than 6 months are equal. .

Keywords: Complete atrioventricular canal defect, early primary repair, 6 months of age.

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List of abbreviations

ABG	Arterial Blood Gases
ACT	Activated Clotting Time
ASD	Atrial Septal Defect
AV	AtrioVentricular
AVSD	AtrioVentricular Septal Defect
CAVC	Complete AtrioVentricular Canal
CAVCD	Complete AtrioVentricular Canal Defect
CPB	CardioPulmonary Bypass
DORV	Double-Outlet Right Ventricle
DSCAM	Down Syndrome Cell Adhesion Molecule
DS-CHD	Down Syndrome-Congenital Heart Disease
EET	Endo-Tracheal Tube
ICU	Intensive Care Unit
IVS	Inferior Vena Cava
LIL	Left Inferior Leaflet
LSL	Left Superior Leaflet
LA	Left Atrium
LV	Left Ventricle
LVOT	Left Ventricular Outflow Tract
LVOTO	Left Ventricular Outflow Tract Obstruction
MAC	Maximal Alveolar Concentration
MR	Mitral Regurgitation (regurge)
OR	Odds Ratio

P2	Pulmonary component of 2ND heart sound
PFO	Patent Foramen Ovale
PMs	Papillary Muscles
PVOD	Pulmonary Vascular Obstructive Disease
PVR	Pulmonary Vascular Resistance
RA	Right Atrium
RV	Right Ventricle
SD	Standard Deviation
SVC	Superior Vena Cava
SPAP	Systolic Pulmonary Artery Pressure
TGA	Transposition of the Great Arteries
TOF	Tetralogy Of Fallot
VSD	Ventricular Septal Defect

Introduction

Since the first documented description of Complete Atrio-Ventricular Canal Defect (CAVCD) repair by Lillehei and colleagues in1954, various techniques have been proposed for its repair. *(Mee et al 2002)*

These techniques have been continuously refined due to a better understanding of the surgical anatomy as well as the patho-physiology of systemic -to- pulmonary shunting associated with this defect and the resulting pulmonary vascular obstructive disease (PVOD). *(Yamaki et al 1993)*

However, the ideal timing for the repair remains controversial.

Some authors suggest two-stage repair consisting of pulmonary artery banding as a first stage followed by complete repair thereafter. Never the less, palliation with pulmonary artery banding is confronted by the complications associated with banding as tight band which can cause severe pulmonary stenosis and myocardial hypertrophy as well as loose banding which will end up in unprotected pulmonary vascular bed. *(Najm et al 1997)*

On the other hand, many authors now advocate primary repair at the age of six months or earlier before the onset of irreversible pulmonary hypertension especially in the presence of Down's syndrome. This approach is aided by improved intra-operative myocardial protection and postoperative ICU management in younger infants with CAVCD. *(Hanley et al 1993)*

Aim of work

Our aim in this study is to evaluate the results of early total repair of CAVCD before six months of age with the hypothesis that, with modern techniques, the current risks of CAVSD repair in children younger than 6 months and those older than 6 months are equal.

Review

Chapter 2-A: Definition & Classification

Complete atrioventricular septal defect is a term used to describe a constellation of congenital cardiac structural abnormalities characterized by a defect in the atrioventricular septum as well as atrioventricular valve(s). *(Ramesh et al 2006)*

Based on the extent of the interventricular communication, these defects can be divided into the following three groups:

Partial AV canal defects consist of an ostium primum atrial septal defect (ASD) and two distinct AV valve orifices with a cleft between the left superior and inferior leaflets but without an interventricular communication. This cleft is usually complete though may be partial or absent. Isolated cleft of the anterior leaflet of the mitral valve also occurs but generally would not be categorized as partial AV canal. Down syndrome is rare in this group, occurring in less than 10% of patients. (*Pic.1&5 plate1)*

Transitional AV canal defects consist of an ostium primum ASD, two AV valves with a left-sided cleft and a restrictive ventricular septal defect (VSD). This VSD is usually small and partially closed by the attachments of chordae from the superior and inferior bridging leaflets to the crest of the interventricular septum.

Complete AV canal defects constitute the other end of the spectrum and consist of an ostium primum ASD, one common AV valve orifice and a nonrestrictive VSD in the inlet portion of the interventricular septum. *(Pic.5&6 plate 2)* Moreover, inlet VSD and a cleft mitral valve with a restrictive or absent ASD should be considered to be within the spectrum of AVCdefects. *(Jacobs et al 2000)*

Rastelli classification for Complete AVC defects. *(Fig.1)*

In 1966 Rastelli and Kirklin described a classification of CAVC defects based on the extent of bridging of the left superior leaflet (LSL) across the interventricular septum. [The left inferior leaflet (LIL) is usually not well developed being short and immobile with rolled edges, and also it displays greater anatomical variation.] Although it is an oversimplification and represents categorization of a continuum of leaflet abnormalities, it may have a role in the repair of complete AV canal defects using the *classic single-patch technique* where recognition of whether the bridging leaflets are undivided over the crest of the interventricular septum is important because undivided leaflets need to be surgically incised into right and left components to allow placement of a patch to close the interventricular and interatrial communications. However, the Rastelli classification is of less importance in the *double-patch or modified single-patch techniques* which will be described later. *(Rastelli et al 1966)*

(1) Rastelli A defect: *(pic.2 plate 1)*

It is the most common type which is present in approximately 75% of patients especially in patients with Down syndrome.
In type A there is complete division of the superior common leaflet over the crest of the septum. Therefore there are chordal attachments from the crest of the septum separating the superior common leaflet into left and right septal components with no bridging of the LSL.

(2)Rastelli B defect:

This is a rare type that is often associated with unbalanced AVCD. There is mild to moderate bridging of the LSL. Its chordae attach either from the tricuspid component into the left ventricle (usually seen when there is left ventricular dominance) or from the mitral component to the right of the crest of the inter-ventricular septum or to a prominent papillary muscle in the right ventricle (RV) (usually seen when there is right ventricular dominance).

(3)Rastelli C defect:

This type occurs in Approximately 25% of patients, usualiy in association with other complex heart lesions such as tetralogy of Fallot (TOF), transposition of the great arteries (TGA), and double-outlet right ventricle (DORV). There is extensive bridging of the LSL. Its chordae are free floating and are not attached to the underlying crest of the interventricular septum, but rather attach to the anterolateral papillary muscle of the RV. *(Frank et al 2005)*