Original article:

Characteristical Impact on Surgical approach for repair for Double-Outlet Right Ventricle

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Abstract:

Introduction: By describing a ventriculo-arterial connection rather than a specific congenital malformation, double-outlet right ventricle (DORV) includes a broad spectrum of anatomic variants and associated malformations.

Materials and methods: The study population comprised all patients with a diagnosis of DORV who underwent a surgical procedure between .Patients with complete AV canal defect were excluded.A diagnosis of DORV was made if both great arteries originated predominantly from the right ventricle with application of the "50% rule," which requires that one great artery arises completely and the second >50% from the right ventricle and aorto-mitral continuity.

Results: This study confirms that a biventricular repair can be achieved in the great majority of patients with DORV with an acceptably low perioperative mortality.

Conclusion: The surgical intervention needs to be tailored according to the surgical anatomy.

Introduction:

By describing a ventriculo-arterial connection rather than a specific congenital malformation, double-outlet right ventricle (DORV) includes a broad spectrum of anatomic variants and associated malformations. In 1964, Kirklin et al reported successful correction in a child with DORV, subaortic ventricular septal defect (VSD), and concordant atrioventricular (AV) connections.Since then, complete correction through the use of a variety of surgical techniques has been achieved in more complex forms of DORV. This present study analyses the anatomic findings, surgical strategies, and results among patients with DORV presenting for surgery in a single institution over a 3 year period.

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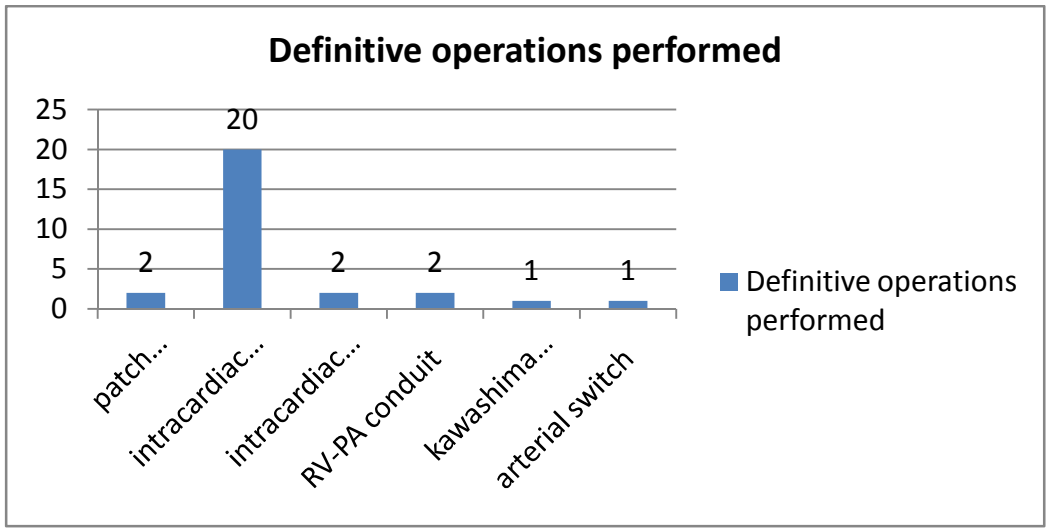
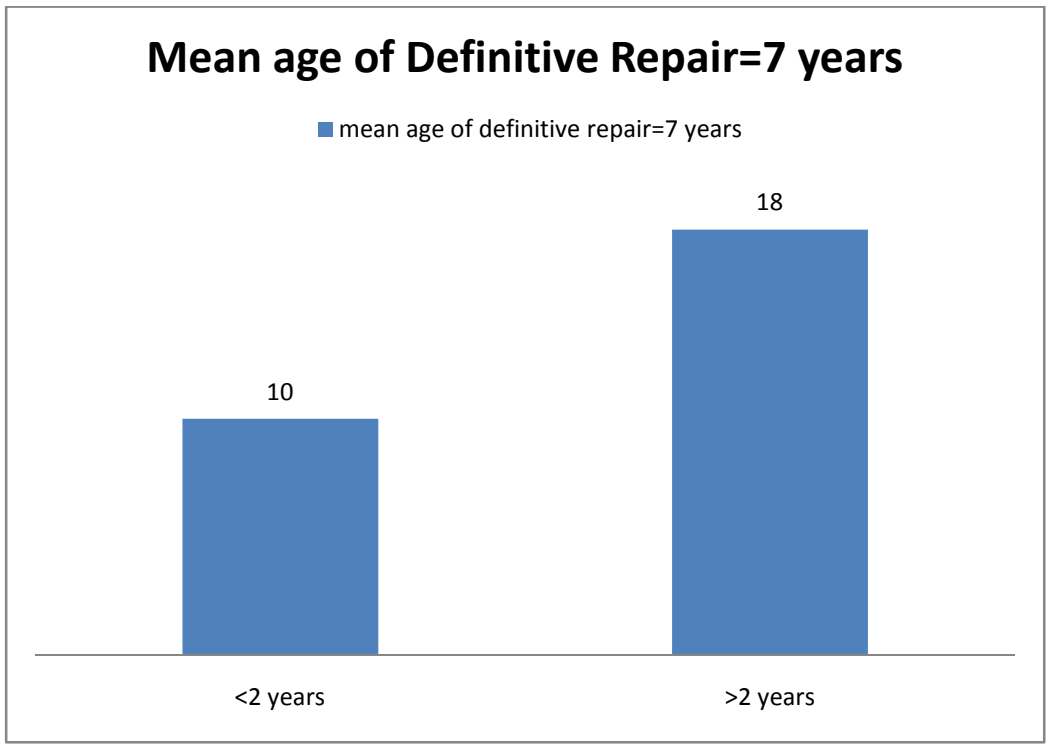
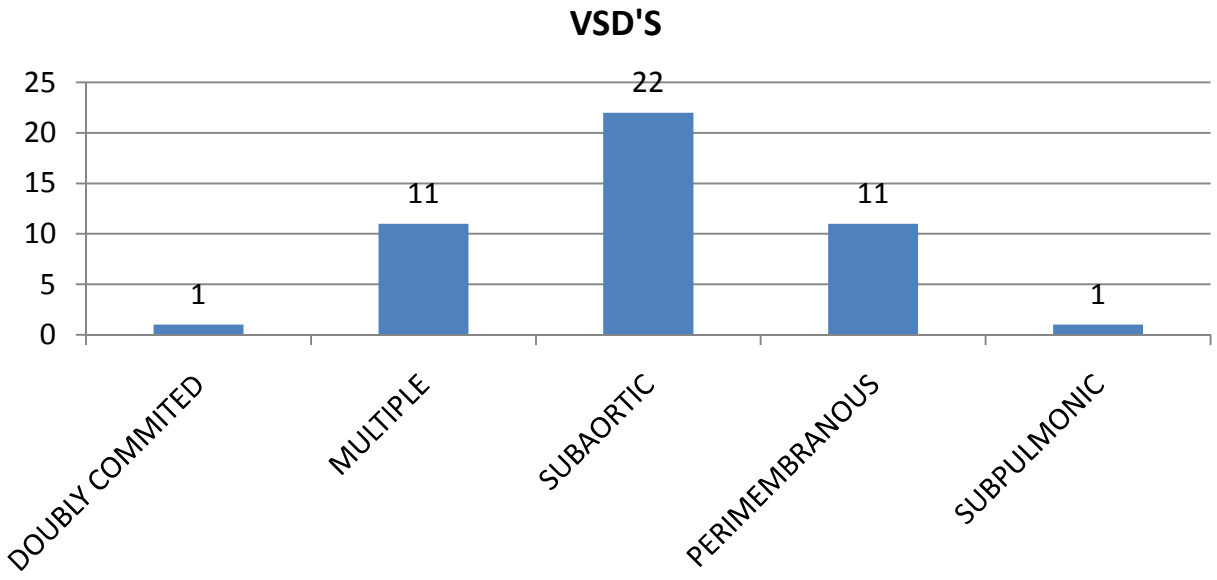

Definitive operations performed
25
20
15
10
5
0
2
20
2
2
1
1
patch...
intracardiac...
intracardiac...
RV-PA conduit
kawashima...
arterial switch
Definitive operations performed


Mean age of Definitive Repair=7 years
mean age of definitive repair=7 years
18
10
<2 years
>2 years

S.NO	SIMPLE ANATOMY	COMPLEX ANATOMY
1	Single VSD	Multiple/Doubly committed VSD
2	Aortic override <100%	Aortic override 100%
3	No major pulmonary artery anomaly	Pulmonary artery anomaly
4	No Associated anomalies	Associated anomalies

Anatomic features:



Pulmonary outflow tract obstruction (POTO):

- Infundibular and valvular PS most common in patients with subaortic VSD's.
- Pulmonary outflow tract obstruction (POTO) was more common in subaortic VSD's.
- Majority of patients with POTO belonged to complex group.
    - LSVC with MAPCA with RV dysfunction = 1
    - Post BT shunt = 1
    - LSVC with narrow MPA = 1

S.NO.	ASSOCIATED ANOMALIES	NUMBER
1	PDA	3
2	LSVC	4
3	RSPV-SVC	1
4	PAPVC/TAPVC	2
5	MAPCA with RV dysfunction	1
6	AV canal defect	1
7	Hypoplastic LV	1
8	Interrupted IVC	1
9	MR with PR	1
10	Accessory cord in LVOT	1
11	Anneurysm of RCC & NCC	1
12	Dextrocardia	1

PRIOR SURGICAL PROCEDURE:

- 4 patients underwent palliative BT shunt.
- Palliative surgeries were performed in complex anatomy patients

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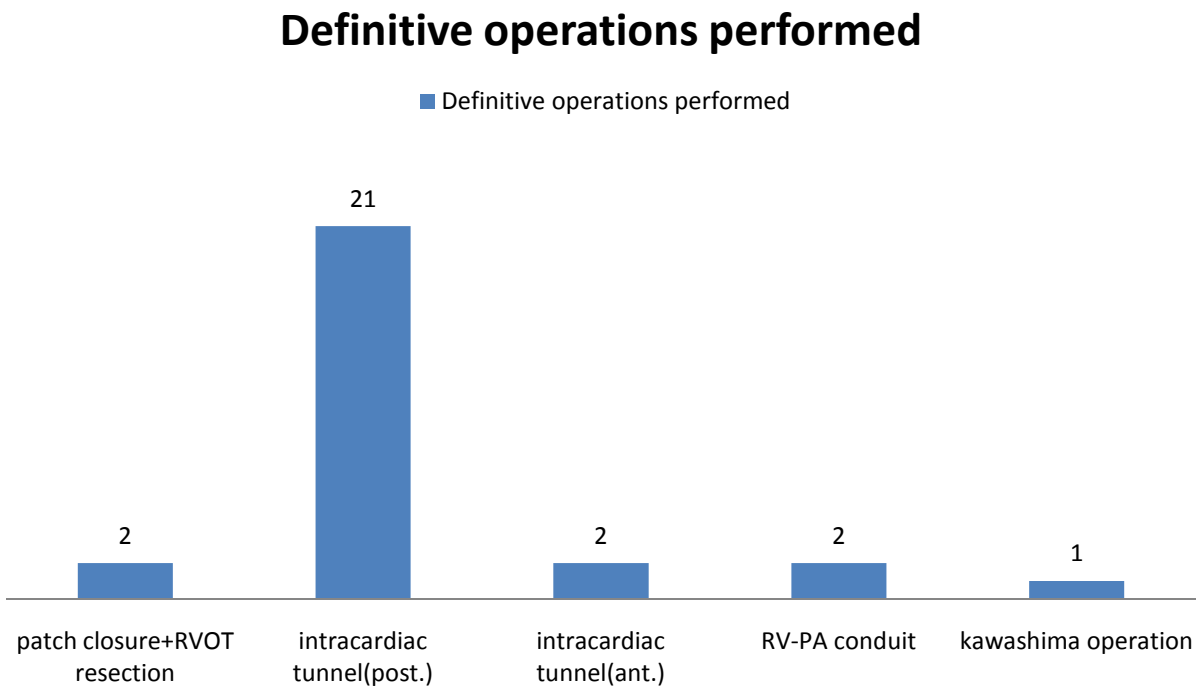
Results:



Surgical outcome

Early mortality:

- Amount of complex anatomy patients died- 3 patients.
- In 2 patients there was difficulty in coming off bypass and patch was perforated.
- 1 patient died after 24 hours had arrhythmias.

This study confirms that a biventricular repair can be achieved in the great majority of patients with DORV with an acceptably low perioperative mortality.

VSD Site and Choice of Surgical Procedure

- Although the classification of DORV by VSD site as introduced by Lev and co-workers remains useful for surgical planning, the present study has not demonstrated a significantly higher risk in patients with non committed or multiple VSD's.
- Among patients with noncomplex forms of DORV and noncommitted VSD, an intra-ventricular baffle repair was still possible in the majority of cases.

- All VSDs in this group were "anatomically" rather than "surgically" noncommitted,with no case in which interposed tension apparatus of the atrio-ventricular valves prevented biventricular repair

RISK FACTORS FOR HOSPITAL MORTALITY

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Multiple ventricularseptal defects:

- By multivariate risk factor analysis of all patients surviving to definitive surgery, the presence of complex anatomy was the only factor related to early mortality. Similar results were reported by Aoki et alin an analysis of patients with DORV undergoing biventricular repair.
- In most cases the presence of additional VSDs does not preclude successful biventricular repair. However, complex forms of DORV remain a challenge, and the optimal surgical approach needs to be individualized according to the specific anatomic features present, including the number and site of any additional VSDs.

The advent of newer surgical and interventional catheter techniquesmay further facilitate biventricular repair and improve the results in this group of patients

Early surgical repair:

- Patients who underwent definitive repair before 2 year of age were also at significantly higher risk in the present series, but this group tended to include patients with unfavorable anatomic variants.
- Early definitive repair is seldom required in most patients with DORV and is probably best avoided in patients with complicating anatomic features.

Limitations of the study:

- The classification of varying anatomic DORV subtypes into simple and complex forms has, to some extent, been arbitrary.
- However, this had no bearing on risk factor analysis, in which anatomic variables were considered separately, regardless of whether the anatomy was regarded as complex or noncomplex.
- This study has the inherent limitations of a retrospective review in which patients were not randomly allocated to varying treatment groups.
- Risk factor analysis is, by its nature, predicated on outcome, which may in turn be influenced by varying treatment options.

Conclusion:

From present we may conclude , the surgical procedures in a cohort of consecutive patients with DORV and a wide range of anatomic features have been examined . Early mortality was low among patients with noncomplex forms of DORV undergoing biventricular repair but was higher in patients with complicating anatomic features undergoing similar surgery. Risk factors for early mortality identified from multivariate analysis include complex anatomy and definitive surgical repair at early age. Finally the surgical intervention needs to be tailored according to the surgical anatomy.

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