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Double Outlet Right Ventricle with Non-Committed Ventricular Septal Defect and Other Complex Associated Anomalies in an Infant: Overview of the Surgical Management Approach

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Abstract:- In the first place, the right ventricle is the primary or only point of origin for both main arteries in a patient with double outlet right ventricle (DORV). In order to guarantee proper repair, a wide variety of surgical approaches, considerations, and procedures are required for the wide spectrum of disorders that make up DORV. A functional classification based on physiological characteristics is one technique to classifying it, which might help determine the appropriate surgical strategy. Presentation of the case: Our case study focused on a female newborn infant who was 2 months old when she arrived at the hospital complaining of worsening shortness of breath and anorexia. The patient was diagnosed with pulmonary over-circulation, an abnormal accumulation of blood flowing into the lungs, through the following cardiac abnormalities: double outlet right ventricle (DORV) with a non-committal ventricular septal defect (VSD), no pulmonary stenosis (PS), mitral atresia, atrial septal defect (ASD), single ventricle dominance (SVD), anomalous pulmonary venous drainage (APVD), and patent ductus arteriosus (PDA). After a palliative procedure including the closure of the PDA and the implantation of a PA band, the patient was to have a surgical procedure known as univentricular repair. During the surgical meeting, this course of action was agreed upon. Conclusion: Different atrial configurations, atrioventricular connections, ventricular morphologies, and spatial interactions between the main arteries may all contribute to the wide anatomical spectrum of DORV. In order to decide between biventricular repair and univentricular palliation, several anatomical features must be present in the patient. In addition, palliative treatments may be necessary for certain patients before they may have a permanent surgical repair.

Keywords:- *Double Outlet Right Ventricle (DORV); Univentricular repair; Ventricular Septal Defect (VSD); Pulmonary over-circulation; Palliative treatment.*

I. INTRODUCTION

The traditional definition of Double Outlet Right Ventricle (DORV) is that DORV is a condition in which both of the major arteries originate mostly or entirely from the right ventricle. This description purposefully embraces a broad spectrum of situations. Around three to nine cases are reported for every one hundred thousand live births. Along with other severe congenital heart defects (CHD), DORV may present itself as a cardiac aberration that is independent of any other heart issue, it can coexist with other heart disorders, or it can be accompanied by extracardiac abnormalities. The position of the Ventricular Septal Defect (VSD) and the presence or absence of Pulmonary Stenosis (PS) are the primary factors that determine the etiology and clinical symptoms of DORV (Obler et al, 2008; Yim et al, 2018). This affects both the clinical manifestations and the etiology of DORV.

Hearts that have DORV display a broad variety of changes in terms of their physical properties, connections, and interconnections at various levels of the heart's segments and junctions between segments. These differences may be seen in the heart's physical characteristics. As a result, the clinical symptoms and the surgical treatments that are required for people who have DORV may differ. In addition, prior to undertaking a definitive surgical restoration, it is possible that some patients may need palliative therapies (Bell-Cheddar et al, 2023).

Having a non-committed ventricular septal defect (VSD) in conjunction with a Double Outlet Right Ventricle (DORV) is a difficult congenital condition that needs careful surgical treatment. DORV is defined by both great arteries emerging wholly or mostly from the right ventricle, which may rise to a variety of related malformations such as pulmonary stenosis, atrial septal defects, and ventricular septal defects (Sheokand et al., 2021; Cavalini et al., 1998). According to Díliz-Nava et al. (2022), the surgical technique for treating DORV coupled with a non-committed ventricular septal defect (VSD) includes the use of treatments such as the bidirectional Glenn (BDG) surgery as the first step of palliation. Furthermore, the success of treating patients with atrioventricular septal

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defect (AVSD) has extended to encompass individuals with dilated obturator resection of the ventricle (DORV) and varied degrees of ventricular hypoplasia (Shuhaiber et al., 2009).

It is essential for surgical planning to have a solid understanding of the anatomical abnormalities and related anomalies that are linked with DORV. Different classifications, such as those developed by the International Association of Thoracic Surgeons and the European Association of Thoracic and Cardiovascular Surgery, classify DORV according on the kind of VSD that is present. This classification helps in the process of diagnosis and in making choices about therapy (Wang & Li, 2022). Barbero-Marcial et al. (1999) and Ishibashi et al. (2005) found that surgical repair of DORV with a non-committed VSD is still difficult to do. This is because there is often a need for considerable expansion of the VSD as well as precise anatomical modifications.

Additionally, the intricacy of DORV extends to its relationship with other abnormalities such as left ventricular outflow obstruction, which may further complicate the surgical approach (Gopalakrishnan et al., 2017). This combination of defects might make the surgical procedure more difficult. According to Walls et al. (2011), the variability of DORV spans a range of anatomical abnormalities, which highlights the need of personalized surgical techniques. In addition, the existence of hypoplasia of the left ventricle in some DORV variations highlights the many anatomical issues that must be taken into account during surgical care (Rao, 2019).

In conclusion, in order to successfully design therapies for the surgical therapy of DORV with a non-committed VSD, it is necessary to have a complete knowledge of the anatomical variations and accompanying abnormalities. When it comes to maximizing outcomes in children who have this complicated congenital heart disease, it is crucial to have individualized surgical procedures that are guided by exact classifications and anatomical considerations.

II. LITERATURE REVIEW

Having a non-committed ventricular septal defect (VSD) in conjunction with a double outlet right ventricle (DORV) is a severe congenital cardiac condition that needs careful surgical treatment. According to Bajolle et al. (2006), the aberrant development of the outflow zone of the heart is often implicated in the development of such abnormalities as in the case of DORV. The correct diagnosis of DORV is of the utmost importance, and paying attention to the morphology of the channel that connects the ventricles is helpful in evaluating the link between the ventricles and the arterial system (Bharucha et al., 2016). According to Shuhaiber et al. (2009), surgical success in the management of individuals with atrioventricular septal defects has expanded to cover concomitant lesions such as DORV. Furthermore, it has been reported that women who have uncorrected DORV have been able to have successful pregnancies, which highlights the need of receiving expert treatment (Sheokand et al as of 2021).

According to Tchervenkov et al. (2006), the surgical correction of complicated instances involving DORV that are linked with defects such as tetralogy of Fallot or common atrioventricular junction provides a substantial difficulty. According to Rao (2019), a significant number of instances of DORV with mitral atresia and other complicated variants are characterized by hypoplasia of the left ventricle. This condition accounts for a significant percentage of individuals. When it comes to uniting hearts using DORV for the purpose of surgical planning, the orientation of the VSD in relation to the major arteries is of the utmost importance (Uemura et al., 2001). Based on the findings of Ishibashi et al. (2005), it has been shown that extensive expansion of non-committed VSDs in DORV may be effectively done, underscoring the significance of individualized surgical techniques.

In conclusion, in order to achieve the best possible results, the surgical therapy of DORV with a non-committed VSD and other complicated related abnormalities calls for a thorough comprehension of the anatomy that lies under the surface, as well as careful consideration of personalized treatment techniques.

III. CASE PRESENTATION

A 2 months old girl infant referred to Wahidin Sudirohusodo Cardiac Center with presentation of worsening breathlessness and poor feeding. Breathlessness was noticed since newborn without history of cyanosis. History of a term pregnancy, spontaneous labor with birth body mass 2.8 kg and birth body length 49 cm, and normal maternal history. The patient was the 3rd child 3 siblings, no family history of congenital heart disease.

On general physical examination, the patient was conscious, ill-looking with dyspnea, no cyanosis was noted. Current body mass 3.5 kg, body height 58 cm. Blood pressure 78/44 mmHg, heart rate 132 beats per minute, respiratory rate 36 bpm, peripheral oxygen saturation of upper and lower extremities was varying in between 86-88% on room air. Chest examination showed prominent subcostal retraction, vesicular breath sound, rales and wheezing were not heard. Cardiac auscultation revealed single first heart sound, normal split of second heart sound with increased intensity of pulmonal component, murmur was not present.

Echocardiogram examination illustrated mesocardiac, situs ambiguous with interrupted inferior vena cava (IVC) suggesting left atrial (LA) isomerism. Both of great arteries arise from right ventricle (RV) with aorta and pulmonary artery (PA) were seen side by side with muscular inlet type ventricular septal defect (VSD) size 8 mm bidirectional shunt and absence of pulmonary stenosis (PS). Mitral atresia with hypoplastic left ventricle (LV) were visualized. Sinus venosus defect (SVD) type Atrial Septal Defect

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(ASD) size 7 mm left to right shunt in which right pulmonary vein (PV) pooled to RA via SVD and left PV pooled to LA. Patent Ductus Arteriosus (PDA) size 3 mm, left to right shunt, Ao-PA pressure gradient 11 mmHg. Trivial Tricuspid Regurgitation (TR) and Mild Pulmonary Regurgitation (PR). Normal LV and RV systolic function. Echocardiogram was concluded as Double Outlet Right Ventricle (DORV) with absence of PS, VSD muscular inlet bidirectional shunt, Malposition of Great Artery, Mitral Atresia, Hypoplastic LV, ASD SVD bidirectional shunt, Partial Anomalous Pulmonary Venous Drainage (PAPVD), PDA L-R shunt.

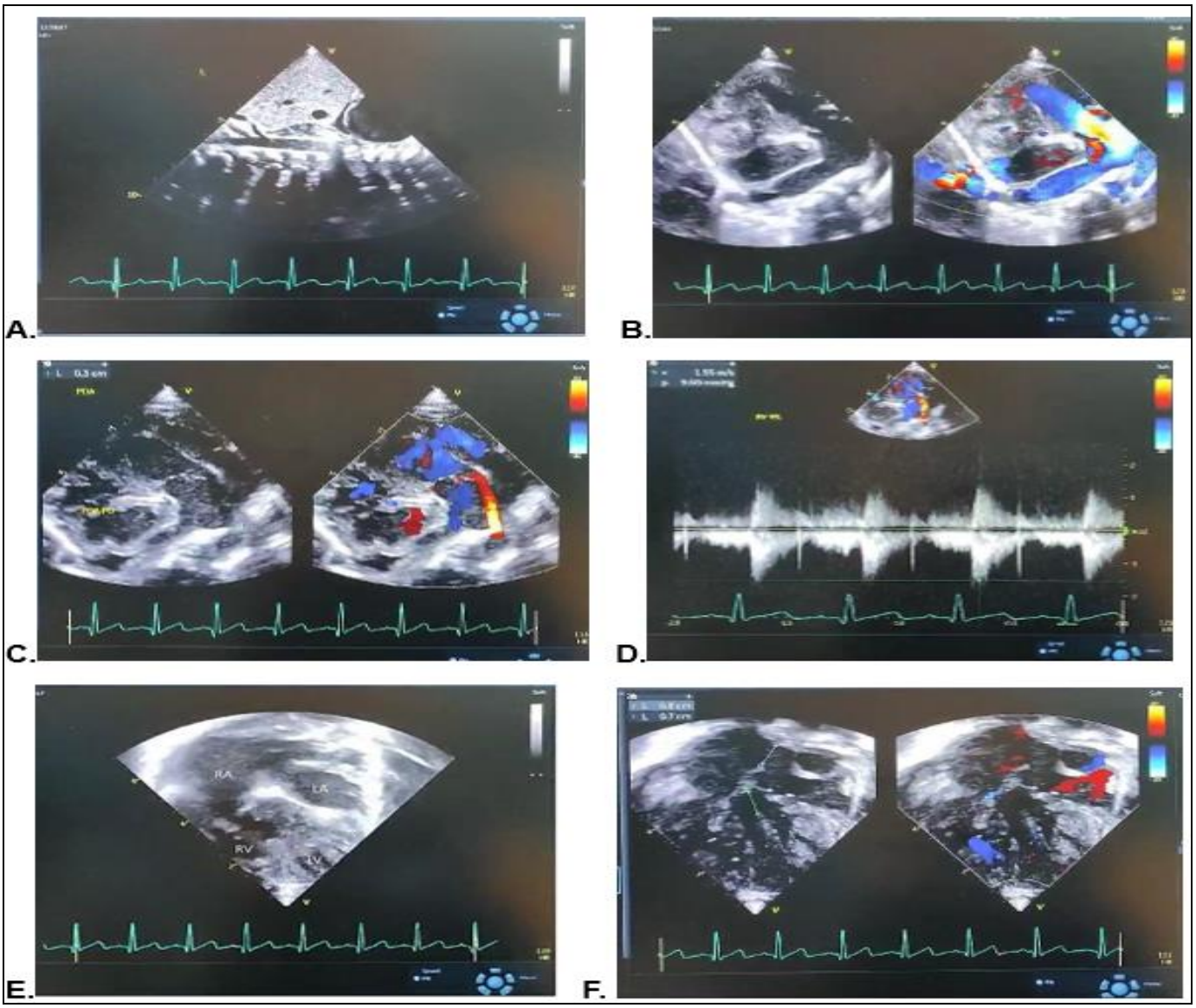

Fig 1: (A) Subcostal View Showed Situs Ambiguous with both Interrupted IVC and Aorta were not Pooled to RA, Suggesting LA Isomerism (B) Parasternal Long Axis (PLAX) View Showed Non-Restrictive VSD and great artery overrides ventricular septum by more than 50%, the great artery has an immediate posterior sweep, suggesting that its is PA, note the separation of anterior mitral leaflet and pulmonary valve (C) Parasternal short axis (PSAX) View Showed Two Great Arteries (Aorta and PA) Were Side by Side, Ao was at the Right Side of PA, PDA was seen with Size 3 mm, L to R shunt, (D) Color Wave Doppler of PA Showed RV-PA Gradient 9.6 mmHg Indicating Absence of PS (E) Apical Four Chambers (A4C) View at Diastolic Phase Showed Hypoplastic LV and Mitral Atresia, (E) Measurement of VSD Muscular Inlet Type Size 8 mm and ASD SVD Type Size 7 mm.

Computed Tomography (CT) Cardiac was performed to validate the echocardiogram finding, which concluded mesocardiac with apex at anterior sinistra, situs ambiguous with RA isomerism, DORV with muscular inlet type VSD with absence of PS, malposition of great artery, mitral atresia, SVD, all PVs to common PV pooled at RA, PDA, and right aortic arch.

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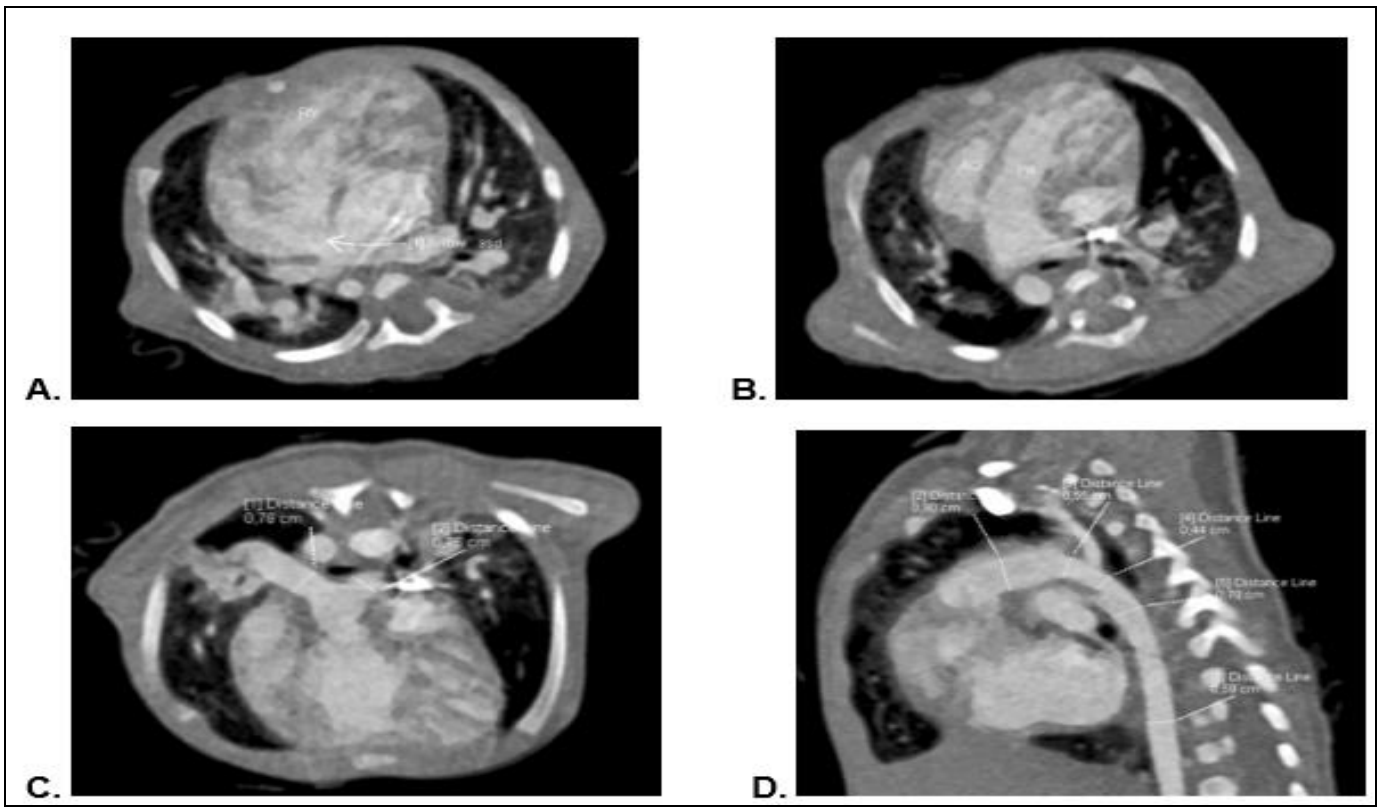

Fig 2: (A) The arrow indicating SVD type ASD, with right and left PV pooled to RA, (B) Side by side great arteries arise from RV, Aorta at the right side of PA, (C) Confluence PA with measurement of proximal RPA 7.8 mm and proximal LPA 7.6 mm, (D) The arrow indicating PDA.

Patient was managed conservatively on oral diuretic (Furosemide 4 mg o.d PO) and ace-inhibitor (Captopril 2 mg t.i.d PO) then referred to surgical conference with result planned for PA banding and PDA ligation subsequently Bidirectional Cavo Pulmonary Shunt (BCPS).

IV. DISCUSSION

The classification of ventricular outflow tract ventricle (DORV) is primarily determined by the precise location of the ventricular septal defect (VSD), the arrangement of the great arteries, the connection between the great arteries, and the presence or absence of obstruction in the outflow tract (Bell-Cheddar et al, 2023; Zamora et al, 1975; Weir et al, 1978). The case that was described was categorized as a non-committed or distant ventricular septal defect (VSD), and the aorta was situated on the right side. Additionally, the patient had a Type III double outlet right ventricle (DORV) with conotruncal abnormalities. Additionally, the patient had abnormalities of both the atrioventricular valve and the ventricle, in addition to a DORV-non-committed VSD that did not include pulmonary stenosis occurring in the patient.

Table 1: Classification of DORV based on various categories

Subaortic VSD	
Subpulmonary VSD	
Doubly commited VSD	
DORV catagories based on great vessel relationshio	
Right anterior aorta	Lef anterior aorta
Right posterior aorta	Left posterior aorta
Right lateral/side-by-side aorta	
DORV categories by Van Praagh	
Type I DORV as an isolated contractual anomali	
Type II DORV with contractual anomalies and associated malsoemacticoes of the atrioventricular valve(s) ventricles	
Type II DORV associated with heterotaxy	
Functional catagories by congenital heart surgery nomenclature and database project	
DORV, VSD type (DORV-VSD)	
DORV with subaortic or doubly commited VSD and pulmonary stenosis, Fallot type (DORV-Fallot)	
DORV, with subpulmonary VSD, transposition type (DORV-TGA)	
DORV, non-commited VSD (ne-VSD)	

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When the ventricular septal defect (VSD) is positioned at a distance from both semilunar valves, a condition known as a noncommitted VSD, and there is no pulmonary stenosis (PS), there is an increase in the amount of blood that flows into the lungs. According to Obler et al. (2008) and Yim et al. (2018), this particular kind of atrial isomerism shows up rather often. The investigation of further abnormalities is of the utmost importance. These abnormalities include several types of heart defects, such as Atrial Septal Defect (ASD) or Patent Ductus Arteriosus (PDA), which cause blood to be redirected from the left side of the heart to the right side of the heart. In addition to other heart abnormalities, such as coarctation of the aorta (CoA), severe mitral malformations like mitral atresia, laterality defects like heterotaxy syndrome or isomerism of the atrial appendages, aortic and pulmonary stenosis, interrupted aortic arch (IAA), and aortic arch obstruction like hypoplasia or atresia of a portion of the aortic arch, DORV (double outlet right ventricle) can occur in conjunction with other heart abnormalities. The bicuspid aortic valve, the double inlet-double outlet right ventricle, and the straddling tricuspid valve are only some of the many anomalies that are associated with this condition. The condition known as DORV with hypoplastic left ventricular (LV) is a variant of hypoplastic left heart syndrome. There is a lack of widespread recognition about the notion of simultaneous blocking of the left atrioventricular (AV) link and the systemic outflow tract (Bell-Cheddar et al., 2023). A number of related abnormalities were present in the patient in this particular instance. These abnormalities included left atrial isomerism, mitral atresia, hypoplastic left ventricle, atrial septal defect of the single ventricle type with aberrant pulmonary venous drainage, and patent ductus arteriosus. The presence of these cardiac abnormalities leads to an increase in the amount of blood that flows to the lungs, which ultimately results in a medical disease that is more commonly known as pulmonary over-circulation. The existence of just one ventricle that is capable of functioning is the defining characteristic of this illness from a physiological point of view.

There are many different illnesses that fall under the category of DORV, and in order to successfully treat them, a vast range of surgical techniques, considerations, and procedures are required. Biventricular repairs, such as intraventricular rerouting patch, Rastelli-type repair (with RV to PA conduit), root translocations, and the arterial switch operation (ASO), are included in the spectrum of surgical procedures. Additionally, univentricular staged palliation procedures are included in this spectrum (Bell-Cheddar et al, 2023; Goo, 2021). The key factors that are clinically and surgically significant in patients with DORV include the following: the position of the AV conduction axis in relation to the edge of the VSD, the location of the VSD in the ventricular septum as seen from the right ventricle, the spatial relationship between the VSD and the annulus along the septal leaflet of the tricuspid valve, the size of the VSD, the presence and extent of the muscular infundibulum, the orientation of the conal septum in relation to the edge of the VSD, the relationship between the great arteries, the presence or absence of obstruction in the aortic or pulmonary outflow tracts, the relationship between the VSD and the arterial valves, the specific type of DORV, any abnormalities in the AV valves, the size of the ventricular cavi According to Yim et al. (2018) and Bell-Cheddar et al. (2023), each of these anatomical characteristics has a role in determining whether or not a patient is qualified to undergo a biventricular repair or a univentricular palliation. Additionally, prior to obtaining a permanent surgical repair, some individuals may need palliative therapies in order to alleviate their symptoms. Bell-Cheddar et al., 2023; Vimala et al., 2013; Myung & Mehrdad, 2021) These interventions may involve the placement of a PA band to address pulmonary over-circulation conditions, the insertion of a PDA stent, or the creation of an operative central shunt to address insufficient pulmonary blood flow. These interventions may also involve the creation of an operative central shunt.

The patient's hemodynamics have a significant impact on the treatment that is provided to patients prior to surgical procedures. Diuresis, for instance, is required in situations when there is pulmonary excess circulation since the clinical signs indicate that it is essential. Because of this, there is a possibility that sometimes the individual may need help with breathing as well as the placement of a breathing tube. When dealing with patients who have restricted blood supply to the lungs, it is recommended to take into consideration the possibility of administering prostaglandin E1 infusion and/or atrial septostomy. It is possible that some patients may initially have enough pulmonary blood flow, and they will be able to be released so that they can continue to develop while they wait for surgical intervention. When dealing with a patient who is suffering desaturation and exhibiting indicators of an impaired cardiac output state, it is essential to take into consideration the urgent need for atrial septostomy (Bell-Cheddar et al., 2023). In the example that was reported, there was a disease that caused an excessive amount of blood to flow into the lungs, which led to symptoms such as trouble breathing and inward migration of the lower ribs. As a consequence of this, the patient was first given diuretic medicine in order to minimize the accumulation of fluid prior to having surgical surgery.

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Table 2: List of Essential Modifiers of Surgical Anatomy of DORV

Features	Primary	Secondary
Relationship of the atrioventricular conduction axis to the VSD margin	Perimembranous VSD Nonperimembranous VSD Atrioventricular septal defect	
Location of the VSD seen from the right ventricle	Predominantly outlet Predominantly inlet Confluent inlet and outlet Predominantly apical trabecular Confluent involving all 3 parts	Relationship of the VSD to the tricuspid valve annulus: Along <upper 1/3 Along upper 1/3 to 2/3 Along >upper 2/3
Size and multiplicity of the VSD	Unrestrictive Restrictive No identifiable VSD	Single Multiple
Orientation of the outlet septum relative to the VSD margin	To the left margin of the VSD To the right margin of the VSD Parallel with the plane of the VSD Not related to the VSD margin Deficient or vestigial	
Muscular infundibulum	Subaortic Subpulmonary Bilateral Bilaterally deficient	Extent of the muscular infundibulum Long Short
Great arterial relationship	Normally related Mirror-image of normal Dextro-malposed Levo-malposed Side-by-side with aorta on the right Side-by-side with aorta on the left	
Outflow tract stenosis	Subaortic stenosis Aortic valvar stenosis Subpulmonary stenosis Pulmonary valvar stenosis Pulmonary valvar atresia	Aortic arch Unobstructed Tubular hypoplasia Coarctation Interruption
Type of DORV	VSD location per Lev et al's[9] classification: Subaortic Subpulmonary Doubly committed Noncommitted or remote Aligned with the subaortic outflow Aligned with the subpulmonary outflow Aligned with neither outflow	STS-EACTS-AEPC class: VSD type Tetralogy type TGA type Noncommitted VSD type AVSD
Atriovenricular valve abnormalities	Stenosis of the tricuspid or mitral valve Straddling or over-riding of the tricuspid or mitral valve Insertion of the atrioventricular valve tension apparatus to the margin of the VSD or outlet septum	
Ventricular volumes	Right ventricular volume Enough space for intraventricular baffling Too little space for intraventricular baffling	Left ventricular volume Normal Borderline hypoplasia Too small
Other findings and associated abnormalities	Anomalous systemic venous connection Anomalous pulmonary venous connection Juxtaposition of the atrial appendages Coronary arterial origins and distribution	

Table 3: Criteria for Single or Biventricular Repair of DORV

Univentricular	Biventricular
Small/atretic atrioventricular valve, straddling atrioventricular valve	Normal/nearly normal atrioventricular valve
Remote VSD	VSD that can be incorporated into a baffle
Hypoplastic ventricle	Normal ventricular size
Abnormal coronary anatomy	Conductive coronary anatomy

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The instance at hand exhibits an atretic AV valve (mitral atresia), a distant VSD (muscular inlet VSD), and a hypoplastic ventricle (LV hypoplastic), thereby fulfilling the requirements for univentricular repair of DORV. The patient exhibited a pulmonary over-circulation condition caused by the lack of pulmonary stenosis, abnormal draining of pulmonary veins, underdeveloped left ventricle with mitral atresia, and patent ductus arteriosus. If a single ventricle route is chosen, excessive blood flow to the lungs is first managed by placing a pulmonary artery band (PAB) until the child is 3-6 months old. After these first attempts, a bidirectional cavopulmonary shunt (BCPS) is established. The third step of palliation is accomplished by performing a total cavopulmonary connection (TCPC) when the child reaches 2-3 years of age (Bell-Cheddar et al., 2023; Vimala et al., 2013; Myung & Mehrdad, 2021). Patients with poor pulmonary vascular resistance will eventually experience pulmonary over-circulation. Pulmonary artery banding (PAB) was performed as a preventive measure against the progression of pulmonary vascular disease in individuals experiencing pulmonary arterial overflow. In order to maintain a low pulmonary vascular resistance (PVR) and prevent excessive volume overload on the ventricle, it is necessary to reduce pulmonary blood flow. Pulmonary artery banding (PAB) is preferably carried out before the age of 6 months. This procedure involves narrowing the mid-segment of the main pulmonary artery (MPA), which in turn raises the afterload on the subpulmonic ventricle. As a result, the flow of blood via the pulmonary artery is physically restricted, leading to a decrease in pulmonary blood flow. In patients with single ventricle architecture, the ideal surgical method for pulmonary artery banding is median sternotomy, since it provides optimal visibility of the main pulmonary artery. Patients who often receive preoperative balloon angioplasty (PAB) as part of a phased surgical treatment may have been classified as high-risk candidates for a final repair. According to Bell-Cheddar et al (2023) and Sharma (2012), patients with a mean pulmonary artery (PA) pressure more than 25 mmHg and a resistance greater than 4 wU are not suitable candidates for total cavopulmonary connection (TCPC). In such cases, pulmonary artery banding should be considered as the ultimate palliative treatment.

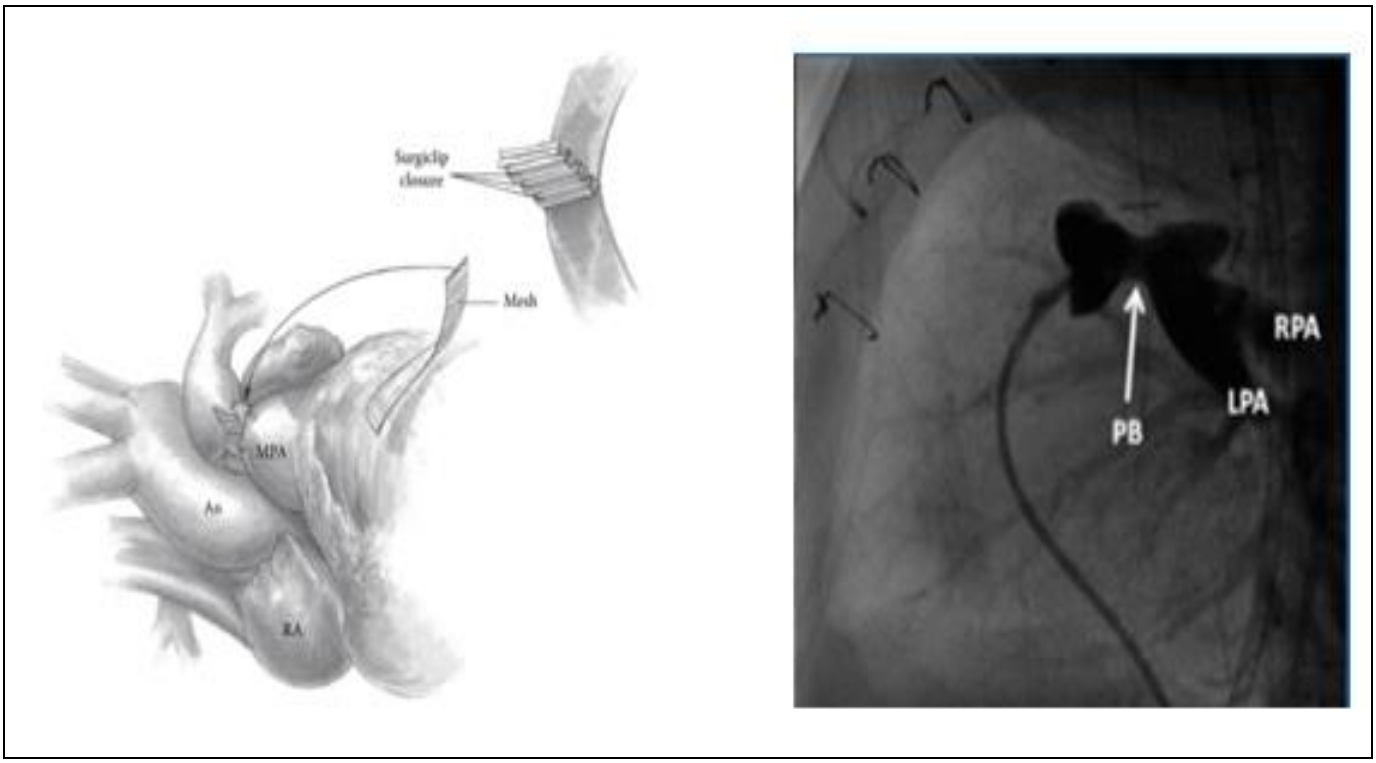

Fig 3: Pulmonary Artery Banding: Pulmonary Blood Flow must be Minimized to Ensure that PVR is Kept Low and the Ventricle does not have an Excessive Volume Load

The second phase of the three-phase management plan for patients with single ventricle physiology serves three purposes: first, to minimize the inefficiencies of the fully mixed circulation as early as possible in life; second, to rectify or eliminate any existing structural abnormalities prior to the Fontan operation; and third, to facilitate ventricular remodeling in response to the sudden decrease in volume load. There are three objectives that are served by the second phase of the plan. Additionally, the second-stage treatment provides the opportunity to treat a number of other anomalies, including branch pulmonary artery hypoplasia or stenosis, arch occlusion, subaortic obstruction, AV valve regurgitation, restrictive atrial septum, or anomalous pulmonary venous connection. These abnormalities may be addressed throughout the surgery. The study that Yasui did in 2009 is the source that is mentioned. When the pulmonary arteries have achieved a size that allows for a low pulmonary vascular resistance (PVR), the best time to do a BCPS is when the pulmonary arteries have reached their full size. This treatment is often

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performed between the ages of four and eight months, or sooner if the child's weight is at least eight to ten kilograms, but it is not performed before three months of age. A mean pulmonary artery pressure of less than 18 mmHg, a Pulmonary Arterial Resistance Index (PARi) of less than 4 wU, and a confluence of the pulmonary artery with a suitable size according to the Kirklin table are the parameters that must be met in order to be eligible for the BCPS procedure. By means of an end-to-side anastomosis, the superior vena cava is linked to the pulmonary artery at the BCPS. In the majority of cases, this can be achieved without the need for any further actions to be taken. On the other hand, an autologous pericardial patch could be used in some circumstances in order to ease any strain that may be placed on the anastomosis or to improve the BCPS. The vena cava superior should, in an ideal scenario, largely direct blood flow towards the right lung rather than the left lung. This is due to the fact that the right lung gets a higher distribution of blood flow. There are times when it can be appropriate to provide the pulmonary artery with an alternate source of additional blood flow instead of the conventional one. Due to the fact that these sources of additional forward flow will assist in the recovery after BCPS, a constricted grouping of the pulmonary artery may be left undisturbed. This will result in a little increase in oxygen saturations. It is possible that it will take many days for the bronchopulmonary circulation system (BCPS) in newborn newborns to properly function and for their oxygen levels to reach an adequate level. It is possible that breathing in nitrous oxide will assist in lowering the pulmonary vascular resistance in the interim. It is essential to effectively treat any remaining cardiac abnormalities throughout the time of BCPS in order to adequately prepare patients for late TCPC. This procedure is performed during the period of BCPS. According to Krishnan (2005) and Hazekamp (2014), an atrial septal connection that is limited ought to be opened when it is required to do so.

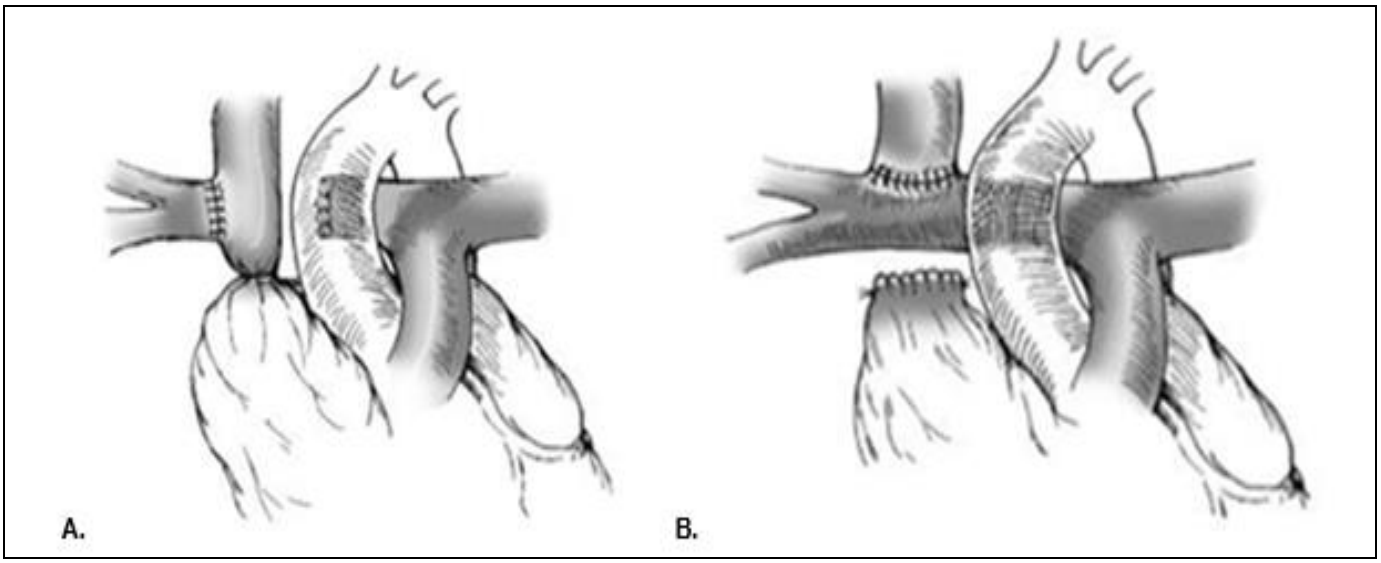

Fig 4: (A) Glenn Shunt. End-to-End Anastomosis Shunt Type from RPA which has been Cut from MPA and Connected with SVC Allowing the Blood to Flow From Proximal SVC towards RPA. SVC Ligated at the Entry Site to RA (B) Modified Glenn Shunt/Bidirectional Glenn Shunt. End-to-Side Anastomosis Shunt Type Between SVC and RA. SVC is Cut from RA and RPA is Not Cut from MPA, Allowing the Blood to Flow Towards RPA and LPA

The TCPC/Fontan approach is the last phase in the palliative treatment process. Its primary objective is to protect the functional single ventricle (FSV) by reducing pressure overload and volume overload, as well as increasing oxygen levels. There are a number of particular requirements that must be satisfied in order to obtain a good result and bring about a reduction in death rates. These include: a low PVR (pulmonary vascular resistance) of less than 3 WU, a low pulmonary artery pressure (PAP) of less than 15 mmHg, an adequate size of the pulmonary artery (PA) in relation to the aorta (PA-Ao diameter ratio greater than 0.75), the absence of significant regurgitation in the AV valves, the presence of sinus rhythm, the candidate being between the ages of 2-4 years or older, previous shunt procedures not having negative effects, normal volume of the right atrium (RA), normal drainage of the caval veins, and the possibility of achieving a three-stage Fontan pathway without the need for cardiopulmonary bypass, which may be safe and feasible in certain patients with a single ventricle and cardiac abnormalities such as DORV (double outlet right ventricle). The procedure known as transcatheter pulmonary valve replacement (TPVR) is normally performed using an extracardiac conduit in the majority of cases. On the other hand, a lateral tunnel type TPVR surgery is carried out in circumstances when it is not feasible to implement a direct passageway from the inferior vena cava (IVC) to the pulmonary artery (PA). It is essential for the extracardiac conduit to have a breadth that is appropriate for adult life in order to satisfy adult life expectations. A Gore-Tex vascular prosthesis of at least 18 or 20 millimeters in diameter is normally required for this. In order to meet this requirement, the child must weigh at least 15 kilograms, which often happens between the ages of three and four years. The fenestration that we utilize is always four millimeters, regardless of the circumstances. Within one to

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three years of the TCPC procedure being performed in the catheterization laboratory, the fenestration will either close on its own at a later time or will be closed spontaneously. While the heart is still beating, transcranial cardiopulmonary bypass (TCPC) is conducted utilizing cardiopulmonary bypass (CBP) with bicaval cannulation at a temperature of 28-30 degrees Celsius. In situations when intracardiac repairs are required, cardioplegia is used to bring the heart to a standstill, and the aorta is only cross-clamped in certain circumstances. It is of the utmost importance to properly position the cannula at an appropriate distance from the atrium and to ensure that the inferior vena cava becomes adequately developed. At the beginning of the procedure, the vena cava is severed from the atrium, and the aperture of the atrium is momentarily narrowed. For the purpose of establishing a link between the Gore-Tex tube and the inferior vena cava, a continuous 6-0 prolene suture provides the necessary support. After that, a hole measuring four millimeters is made in the Gore-Tex tube. After that, the atrial aperture, which is the location where the inferior vena cava is joined, is sewn to the Gore-Tex tube, which covers the hole from the inside. After the tube has been secured, there is no longer any possibility of air entering the heart. During the dissection of the right pulmonary artery, the left atrium is separated from the right atrium to ensure proper anatomy. In the next step, the right pulmonary artery is first properly secured, and then the bottom side of the artery is carefully opened. In the event that clamping becomes difficult, the flow of PB is decreased, and a sucker with a tiny tip is used in order to assure that the PA remains dry. It is common practice to angle the connection between the right pulmonary artery (PA) and the Gore-tex tube in such a manner that it causes a little incline in the direction of the left pulmonary artery (PA). By using 6-0 Prolene sutures, the use of surgical glues is rendered unnecessary. All of the clips on the Gore-Tex tube are removed, and the tube is thoroughly cleaned of unwanted air. In order to eliminate any possible interference with the TCPC's ability to perform its functions correctly, all sources of extrapulmonary flow that are still present have been halted. When using a two-stage strategy, with the first phase being the BCPS procedure followed by the TCPC procedure, and when proper patient selection has been carried out, postoperative care is often not problematic (Krishnan, 2005; Hazekamp, 2014). This is because the BCPS procedure is the first step in the process.

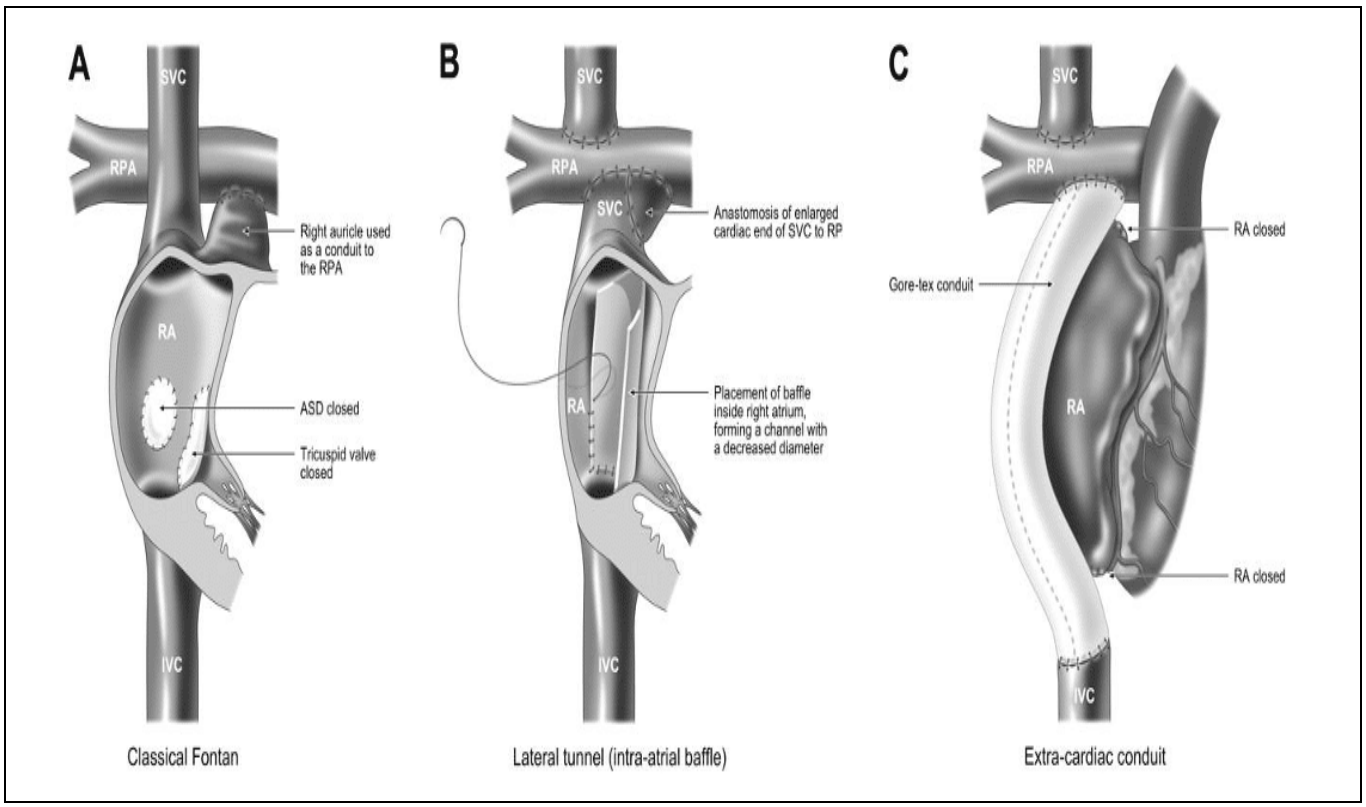

Fig 5: Types of Fontan Procedures

V. CONCLUSION

The structural variations of DORV are highly diverse and may be linked to a variety of factors, including various atrial layouts, atrioventricular connections, ventricular morphologies, and spatial interactions between the main arteries. The existence of these anatomical characteristics is what decides whether a patient is qualified for a biventricular repair or a univentricular palliation to proceed with the procedure. Individuals diagnosed with DORV are subjected to univentricular surgery in situations when biventricular restoration is not feasible, such as when a single ventricle is shown to be functioning normally. An additional point to consider is that before to getting a permanent surgical repair, some patients could receive palliative therapies.

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