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Rheumatic "Acyanotic Taussig-Bing heart" and its Surgical Aspects- A Case Report

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1. Abstract

1.1. Aim: To report a case of acyanotic Taussig-Bing heart, anatomically consistent with L-transposition and rheumatic Left AV valve regurgitation associated with complete AV block in an adolescent male.

1.2. Introduction: Taussig-Bing heart is one of the conotruncal malformation, characterized by double-outlet right ventricle (DORV) and a subpulmonary VSD. Embryologically, abnormal cardiac looping and malalignment of conotruncal septum determines its complexity and great artery relationships.

1.3. Case Report: A 14-year-old acyanotic boy presented with severe left-sided AV valve regurgitation and bradycardia. ECG revealed left sided morphologic right ventricular hypertrophy (RVH) as evidenced by a loss of septal Q waves in left precordial leads suggesting ventricular inversion. X-ray chest revealed a straight upper right cardiac border due to loss of normal relationship of great vessels and cardiomegaly due to both left atrial and morphologic right ventricular enlargements suggesting a left-sided regurgitant lesion. Echocardiography revealed the ventricular inversion, primary origin of both L-transposed great arteries from the left-sided morphologic right ventricle suggesting a 'double-outlet morphologic right ventricle' with 'double discordance' and a subpulmonary VSD of Taussig-Bing type. The left-sided morphologic tricuspid valve is severely regurgitant due to rheumatic process resulting in heart failure which was improved with anti-failure measures and penicillin prophylaxis.

1.4. Conclusion: The presence of left-sided regurgitation associated with bradycardia is a suspicion of 'double discordance' and 'double switch' procedure remains the mainstay of its surgical correction. Left AV valve replacement with intraventricular repair is preferred in this child since the malformation is consistent with

Taussing-Bing type of DORV with L-transposition.

2. Introduction

Taussig-Bing heart" is a morphological subtype of double-outlet right ventricle (DORV) with subpulmonary VSD (ventricular septal defect) [1]. It was first described in 1949 by two outstanding physicians, Helen Brooke Taussig and Richard John Bing, who worked together at the John Hopkins Hospital in Baltimore. The original "Taussig-Bing heart" [2] may be summarized as a double-outlet right ventricle (DORV) with semilunar valves side-by-side and approximately at the same height, a bilateral conus, and a subpulmonary VSD. This original description has been broadened to include all kinds of double-outlet right ventricle with subpulmonary VSD [3],[4]. The morphological variability of "Taussig-Bing heart" is guided by the relationship between great arteries [5] and ventricles that are associated with a bilateral conus [6],[7] and so this case had been reported.

3. Case Report

A 14-year-old acyanotic boy was admitted with features of heart failure in the intensive care unit. His pulse rate was 50 bpm and blood pressure 110/70 mmHg. He had a history of rheumatic fever during childhood and blood chemistry revealed a positive ASO (anti-streptolysin O) titer. Auscultation revealed a grade 4/6 pansystolic murmur over the apex with a shift of apical impulse to left 6th space towards mid- axillary line. ECG revealed high-grade AV block with features of ventricular inversion and strain pattern due to morphologic RVH as shown in (Figures 1 and 2), compared to a normal right-sided pattern as in (Figure 3) with normal echocardiography as in (Figure 4) of a normal adolescent male of similar age. X-ray chest revealed a narrow vascular pedicle with left atrial and morphologic right ventricular enlargement as shown in (Figure 5). Transthoracic echocardiography revealed a VSD with inlet ex-

tension as in (Figure 6) and a discordant relationship between atria and ventricles and a left-sided Ebstenoid morphologic tricuspid valve as shown in (Figure 7) (apical four chamber view), but mRV (morphologic tricuspid valve) is normally attached as in (Figures 8 and 9) (subcostal views). The leaflets are showing the features of rheumatic etiology as in (Figures 10 to 14) and severely regurgitant as shown in (Figures 15 to 18). The child was treated with anti-failure medications such as diuretics, angiotensin-converting enzyme inhibitors (enalapril) with penicillin prophylaxis for a period of one and a half months and the regurgitant lesion became decreased as shown in (Figures 19 and 20) and M-mode mRV study showing the marked improvement in ejection fraction from 36% to 46% as shown in (Figures 21 and 22.) Since the great arteries are primarily originating from morphologic RV as in Figure 23 and it is L-transposed with side-by-side relationship as in (Figure 24) with a subpulmonary VSD, it is consistent with Taussig-Bing anomaly of L-transposition, a Kawashima intraventricular repair was advised if further deterioration of mRV function occurs on periodic follow-up with a continuation of medical treatment along with lifelong penicillin prophylaxis. Palliative procedures are not indicated in this boy since there is no aortic arch obstruction and outflow tract lesions as shown in (Figures 25 and 26).
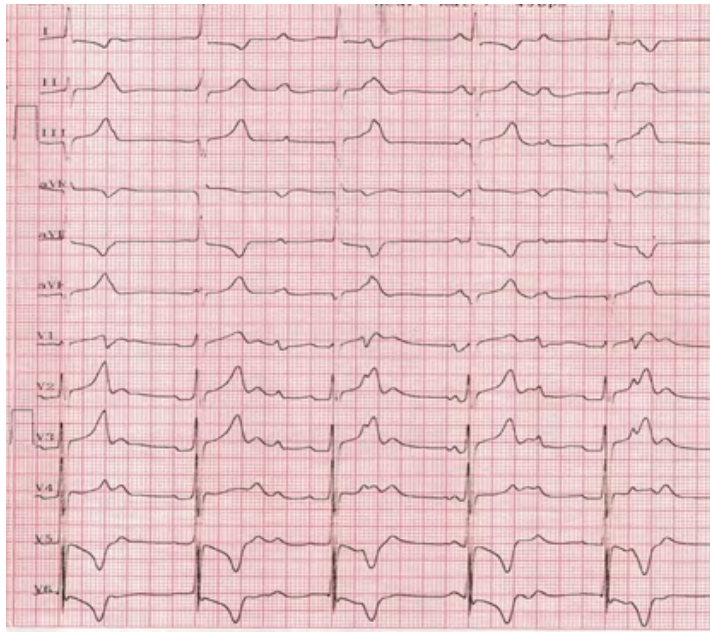

Figure 1: ECG showing complete AV block with narrow QRS (QRS duration < 120 ms) complexes and a heart rate of 50 bpm suggesting a congenital high grade AV block. There is a 'strain' pattern in L1, aVL, V5-V6 with left axis deviation, absence of septal Q waves in left precordial leads due to inversion of bundle branches (reversal of septal depolarization) with a hypertrophied morphologic RV on the left side in a 14-year old boy.
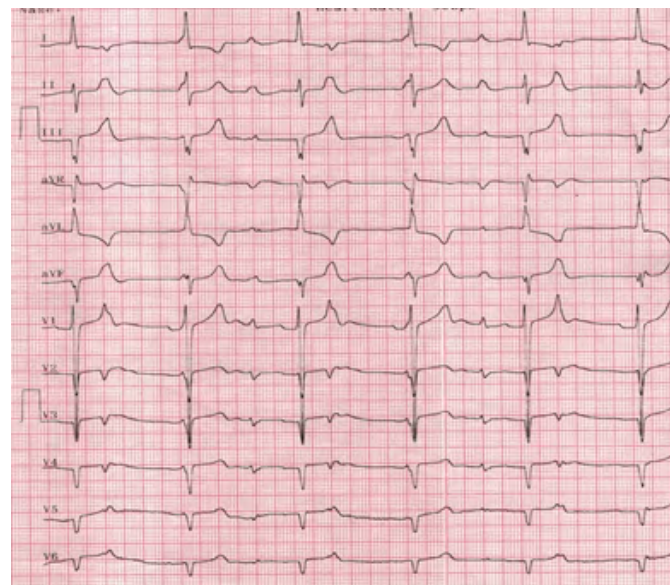

Figure 2. ECG showing the QS complexes in inferior leads (III, aVF) and a QS pattern in right-sided chest leads (V2-V6), compared to a normal ECG of right-sided leads in the same age of a male boy as in Figure 3.
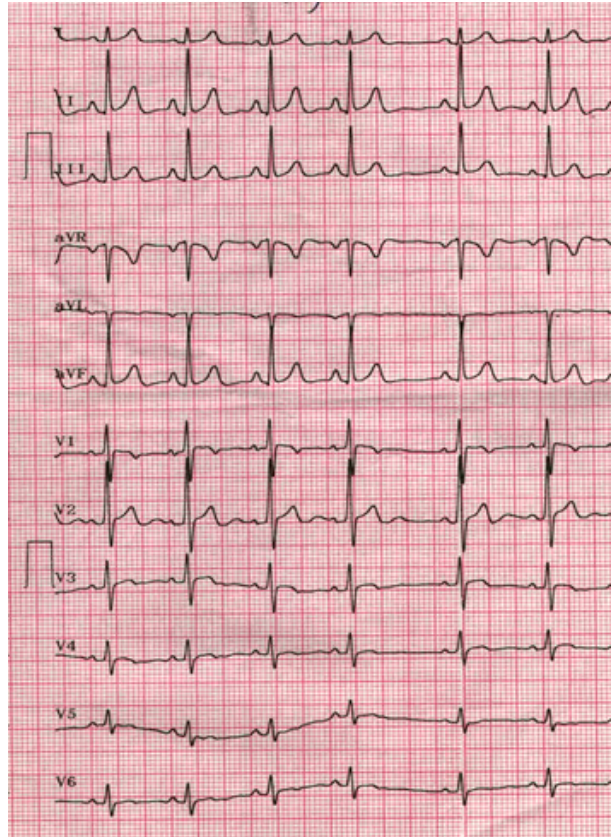
Figure 3: ECG showing RS complexes in the right sided leads of a normal 14-year old boy.

Figure 4: X-ray chest (postero-anterior) view showing the narrow vascular pedicle (right upper arrow) since great arteries are not border forming, appears straight due to loss of normal relationship between arterial trunks as ascending aorta is typically not visualized on the right side, convexities from descending aortic knob and pulmonary trunk are absent on the left side due to posteriorly positioned pulmonary artery suggesting L-transposition. The cardiomegaly is due to aneurysmal Left atrial (LA) enlargement and a dilated morphologic right ventricle (mRV). The visceral situs is normal. Left upper arrow-'hump-shaped' due to inverted RV infundibulum. Left middle arrow- giant left atrium (GLA). Left lower arrow – 'septal notch' corresponding to apical position of interventricular groove.
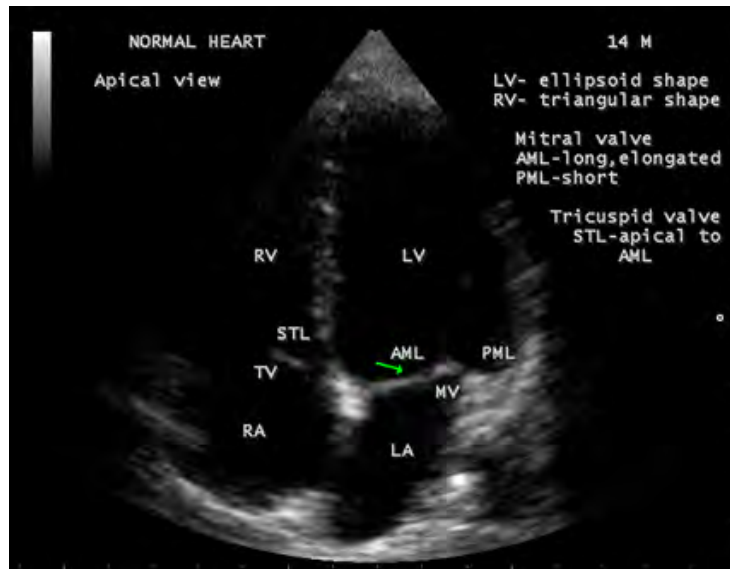
Figure 5: Apical view showing the normal atrioventricular relationships, AV valves (mitral and tricuspid) attachments and the shape of ventricles in a normal 14-year old boy. Arrow- long, elongated anterior mitral leaflet (AML) as a feature of mitral valve.
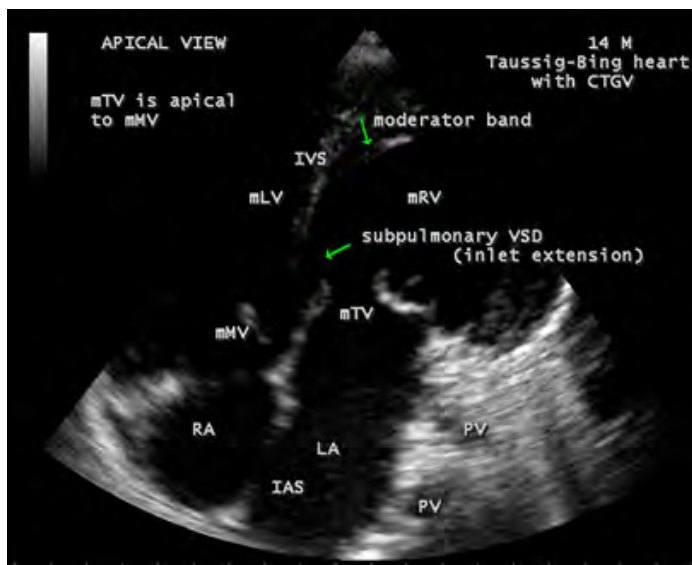

Figure 6: Apical view showing the VSD (ventricular septal defect) with inlet extension. Upper arrow- moderator band. Lower arrow- subpulmonary VSD (ventricular septal defect) with inlet extension.
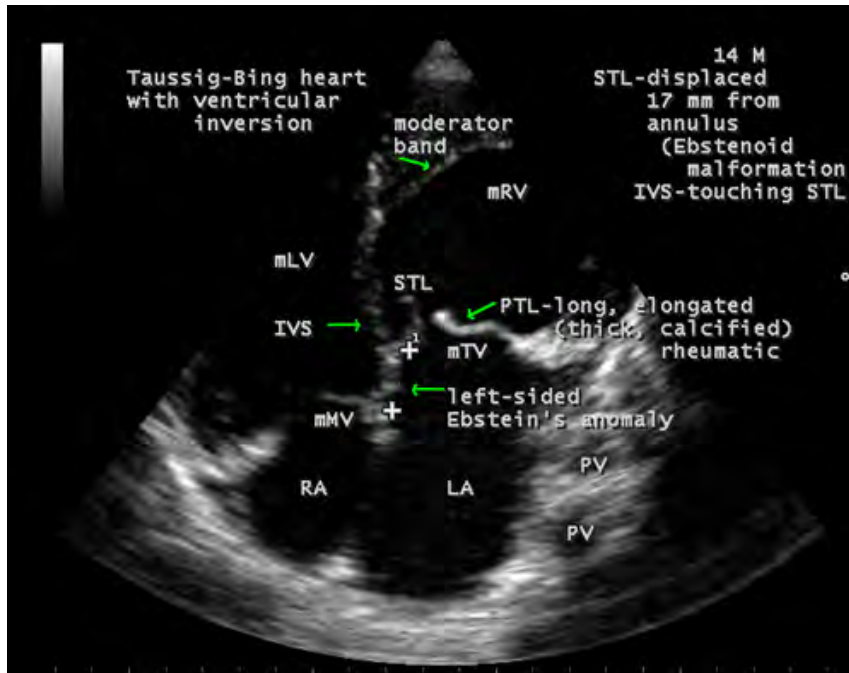
Figure 7: Apical view showing the 'Ebstenoid malformation" of left-sided morphologic tricuspid valve (mTV) due to adherence of septum (IVS) to STL. The mTV is identified by a long and elongated posterior leaflet which is short in mitral valve as in Figure 5 of a normal 14-year old boy and a discordant AV connection. Upper arrow- moderator band. Lower right arrow- IVS (interventricular septum) touching the STL (septal tricuspid leaflet). Left middle arrow- long, elongated posterior tricuspid leaflet (PTL). Lower left arrow- Ebstenoid malformation due to touching of IVS to STL seen as apical displacement of STL and mimicking as Left-sided Ebstein's anomaly.
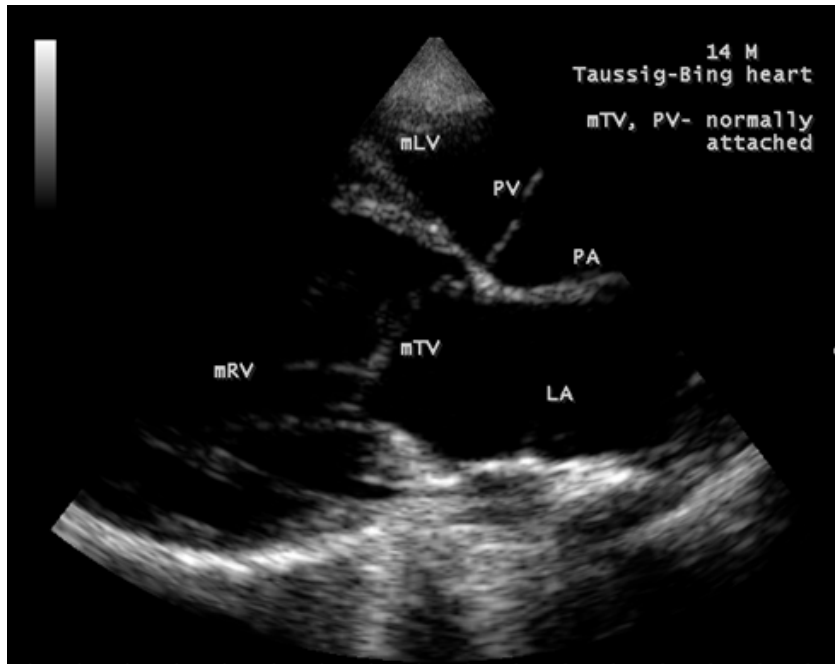

Figure 8: Subcostal view showing the normally attached rheumatic morphologic tricuspid valve and the pulmonary valve.
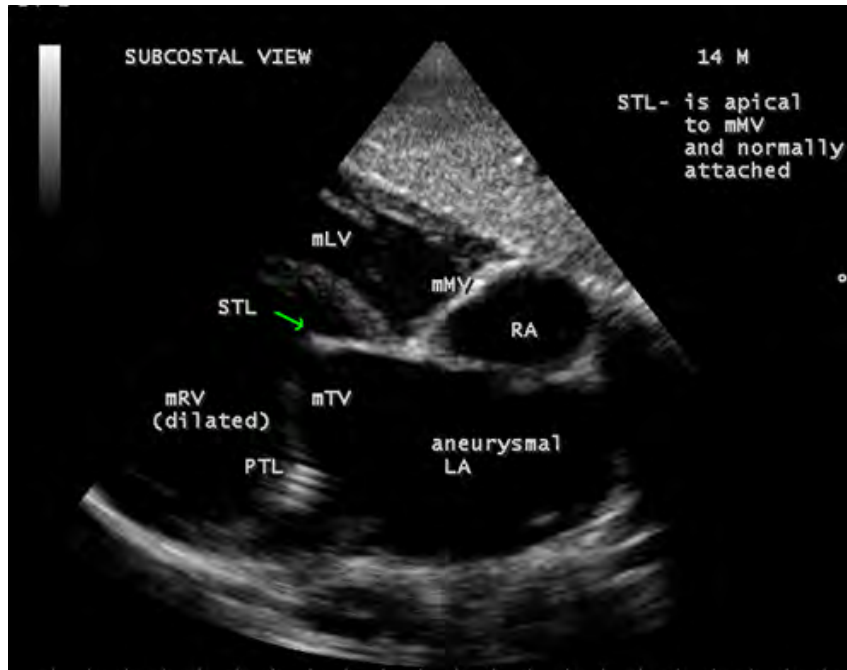
Figure 9: Subcostal view showing the normal attachments of morphologic tricuspid (mTV) and mitral valves (mMV) to the annulus. Arrow- normally attached STL (septal tricuspid leaflet).
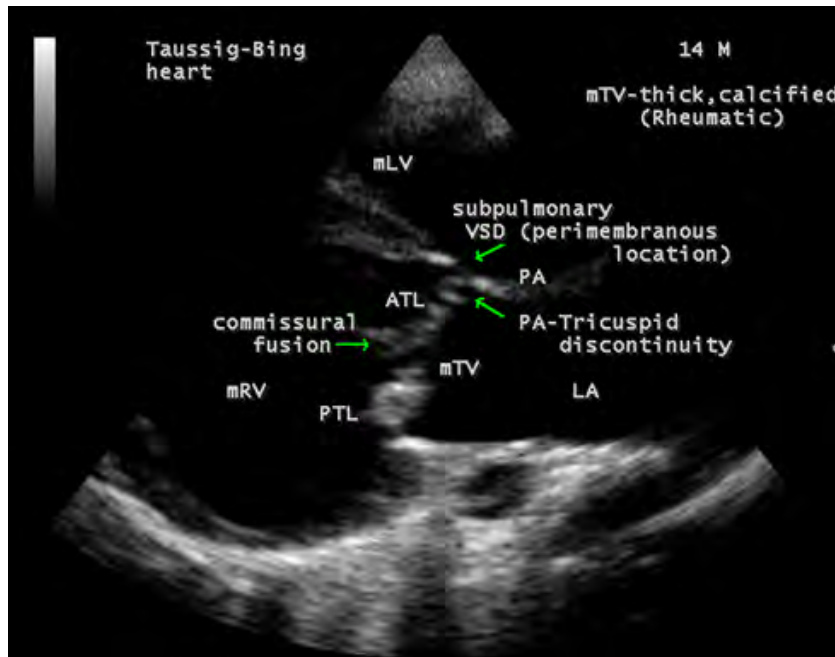
Figure 10: Subcostal view-long sweep –showing the thick, calcified morphologic tricuspid valve (mTV) with commissural fusion due to rheumatic etiology. Upper arrow- subpulmonary VSD (ventricular septal defect) in perimembranous location. Right lower arrow- 'commissural fusion' due to rheumatic process. Left lower arrow- pulmonary to tricuspid discontinuity as a feature of DORV (double-outlet right ventricle).

Figure 11: Subcostal view-showing the calcification and thickening which is more prominent at leaflet tips due to rheumatic involvement, the normal tricuspid annulus (30 mm) and a dilated morphologic right ventricle (mRV) (65 mm)
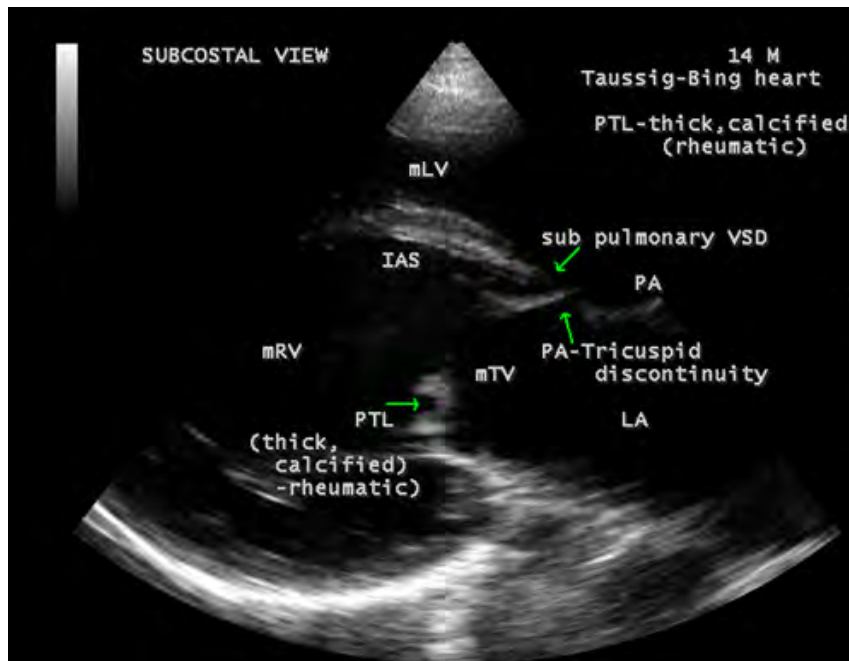

Figure 12: Subcostal view showing the marked rheumatic involvement of posterior leaflet of the morphologic tricuspid valve (mRV). Right arrow-thick, calcified posterior tricuspid leaflet (PTL) due to rheumatic process. Left upper arrow- subpulmonary VSD (ventricular septal defect). Left lower arrow- pulmonary to tricuspid discontinuity as a feature of DORV (double-outlet right ventricle)

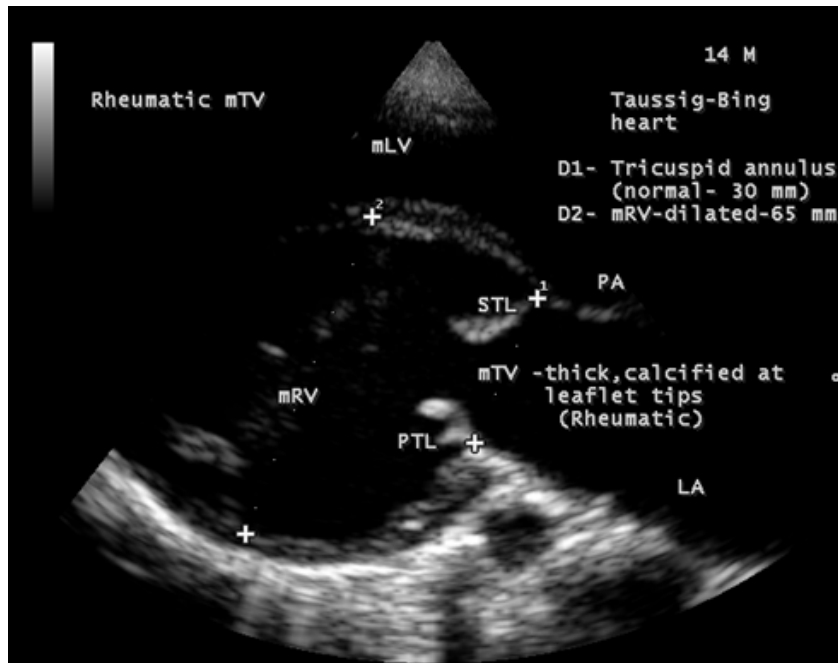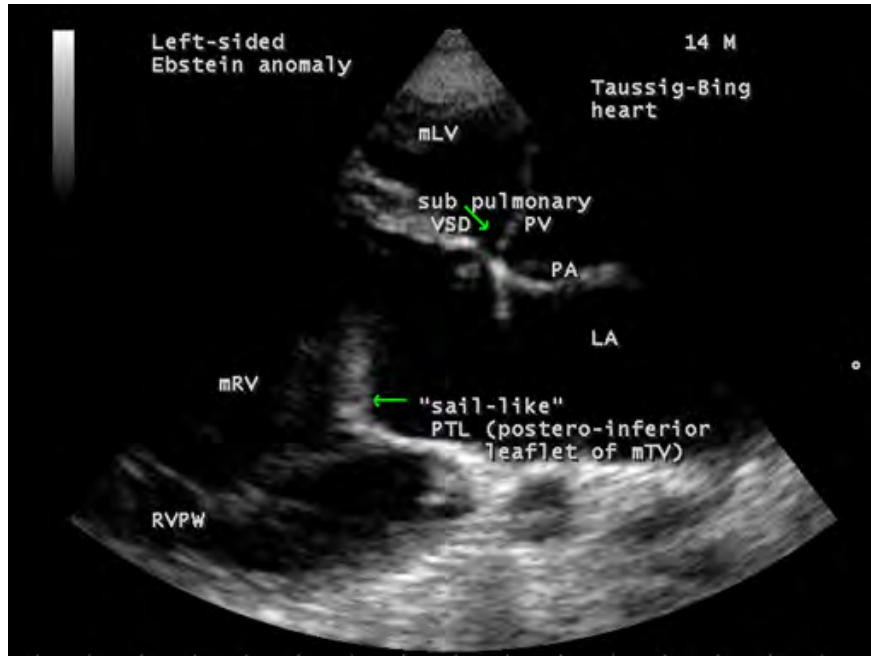**Figure 13:** Subcostal view showing the " sail-like" deformity of posteriot leaflet of morphologic tricuspid valve. Upper arrow- subpulmonary VSD (ventricular septal defect). Lower arrow - "sail-like" posterior tricuspid leaflet.
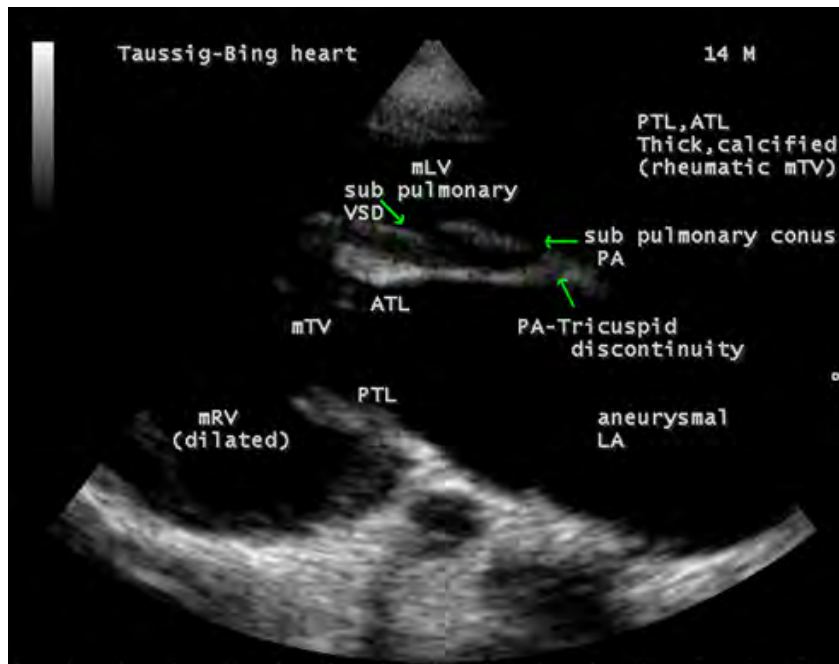
Figure 14: Subcostal view- long sweep showing the subpulmonary conus. Right upper arrow- subpulmonary VSD (ventricular septal defect). Left upper arrow- subpulmonary conus (infundibulum). Left lower arrow- pulmonary to tricuspid discontinuity.
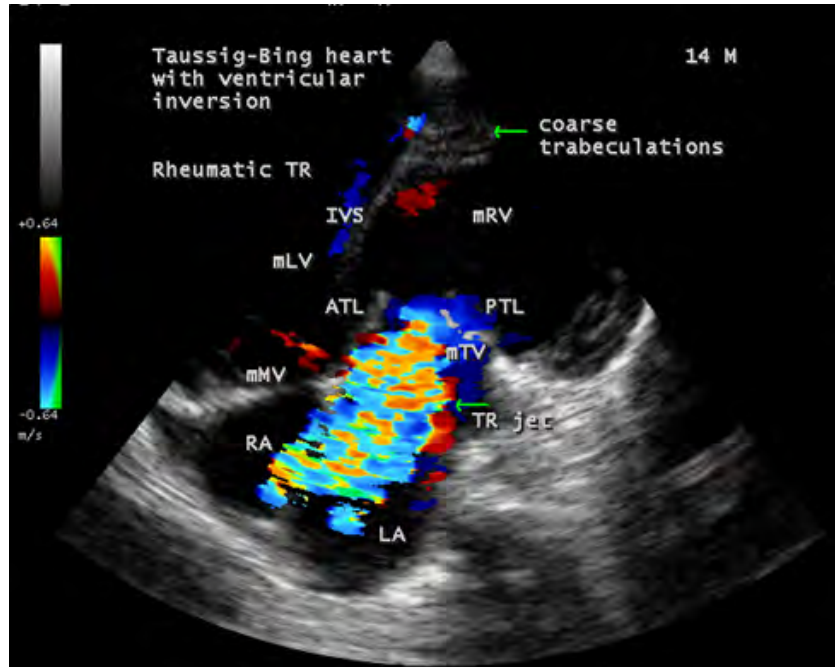
Figure 15: Apical view showing the severe tricuspid regurgitation (TR). Upper arrow- 'coarse trabeculations' as a feature of morphologic right ventricle (mRV). Lower arrow- severe tricuspid regurgitation (TR).
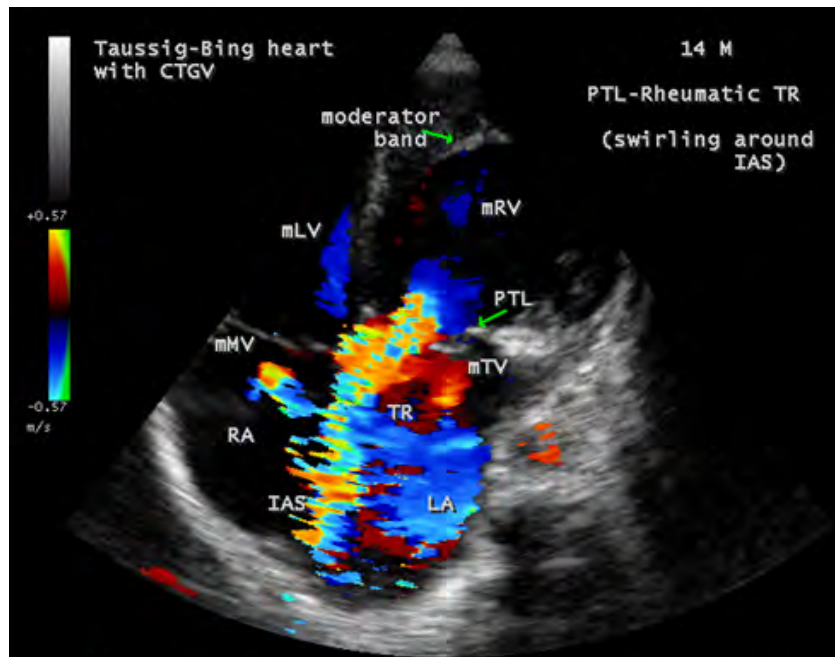
Figure 16: Apical view showing the tricuspid regurgitation jet, originating from the posterior tricuspid leaflet (PTL) swirling around the interatrial septum. Upper arrow- moderator band. Lower arrow- posterior tricuspid leaflet (PTL).
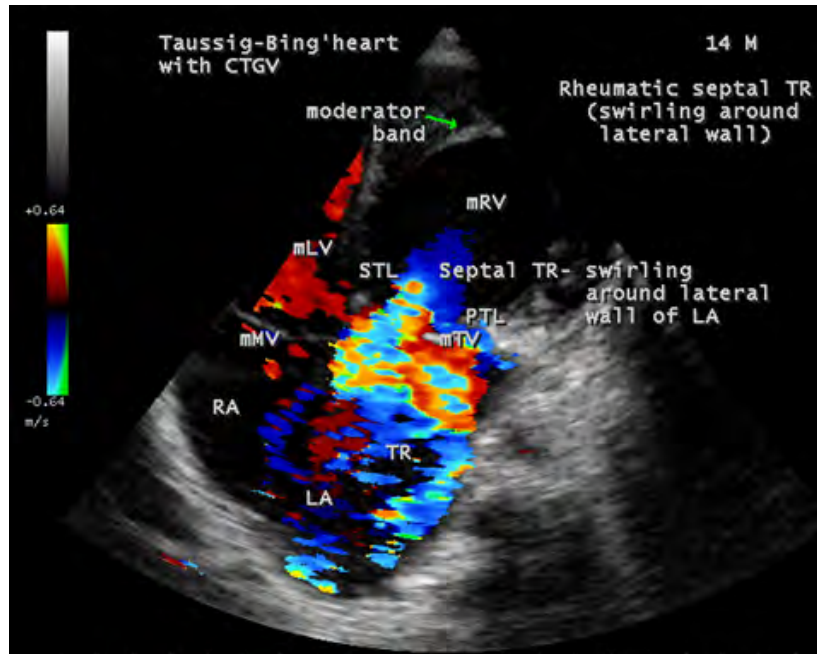
Figure 17: Apical view showing the tricuspid regurgitation jet, originating from the septal tricuspid leaflet (STL) swirling around the lateral wall of left atrium. Upper arrow- moderator band as a feature of morphologic right ventricle (mRV).
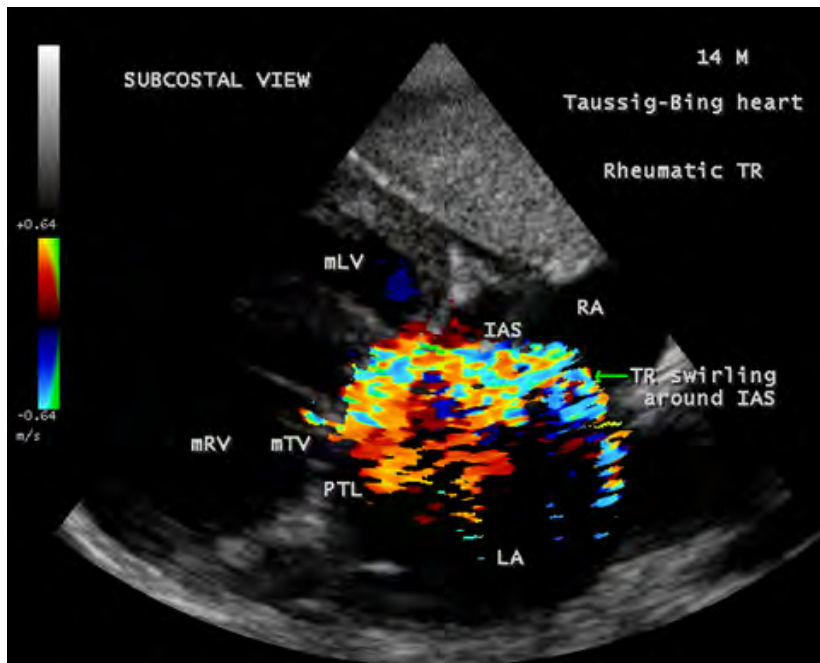
Figure 18: Subcostal view showing the severe tricuspid regurgitation originating from the posterior tricuspid leaflet (PTL) swirling around the interatrial septum. Arrow- Tricuspid regurgitation (TR) .
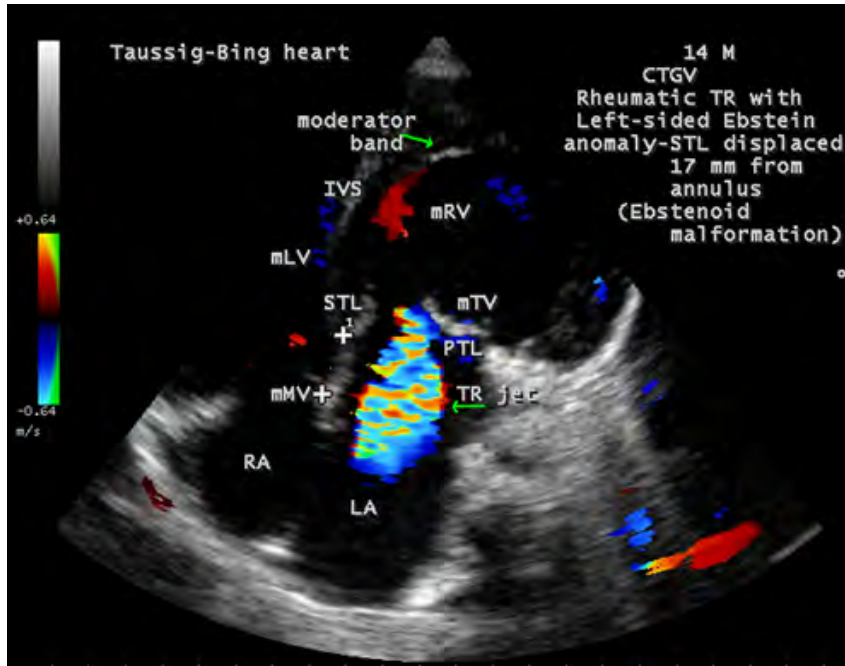
Figure 19: Apical view showing the decrease in regurgitant jet due to aggressive medical treatment and left-sided Ebstenoid malformation due to adherence of septum (IVS) to STL. Upper arrow- moderator band. Lower arrow- tricuspid regurgitation (TR) jet.
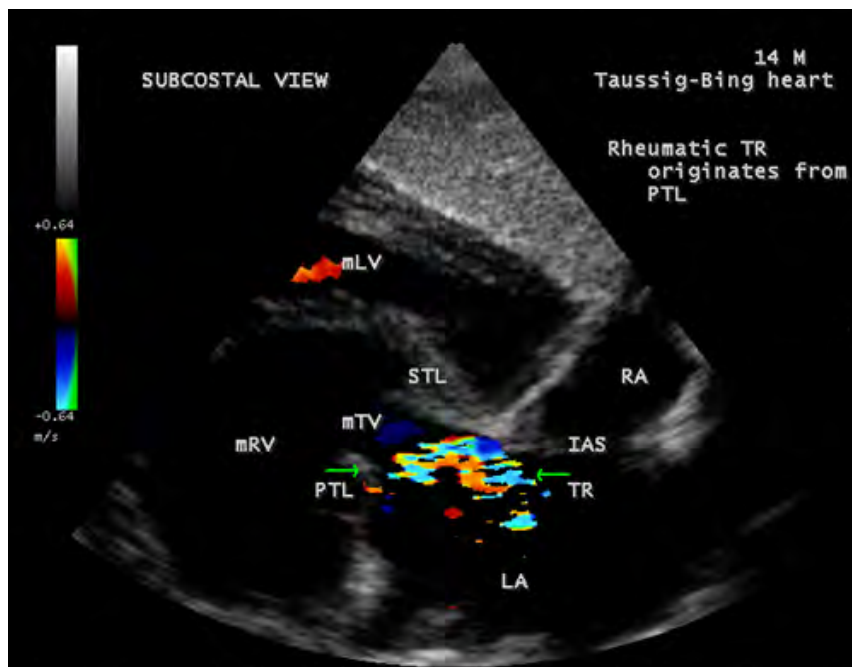
Figure 20: Subcostal view showing the marked decrease in regurgitant jet due to aggressive medical treatment. Right arrow- PTL (posterior tricuspid leaflet). Left arrow- tricuspid regurgitation (TR).
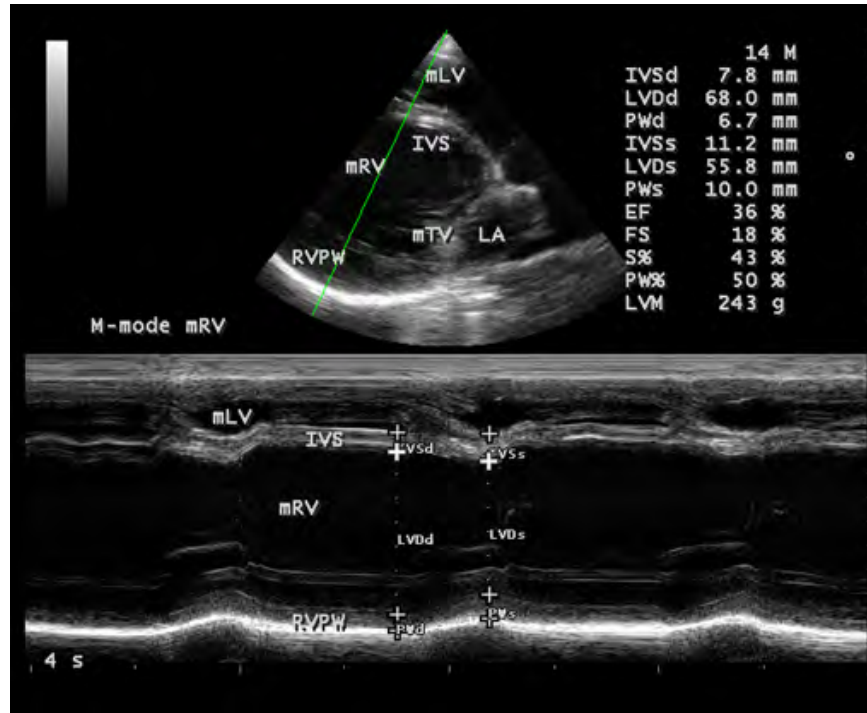
Figure 21: M-mode mRV study (green line) showing the severe dysfunction of morphologic right ventricle (mRV). EF – 36 %.
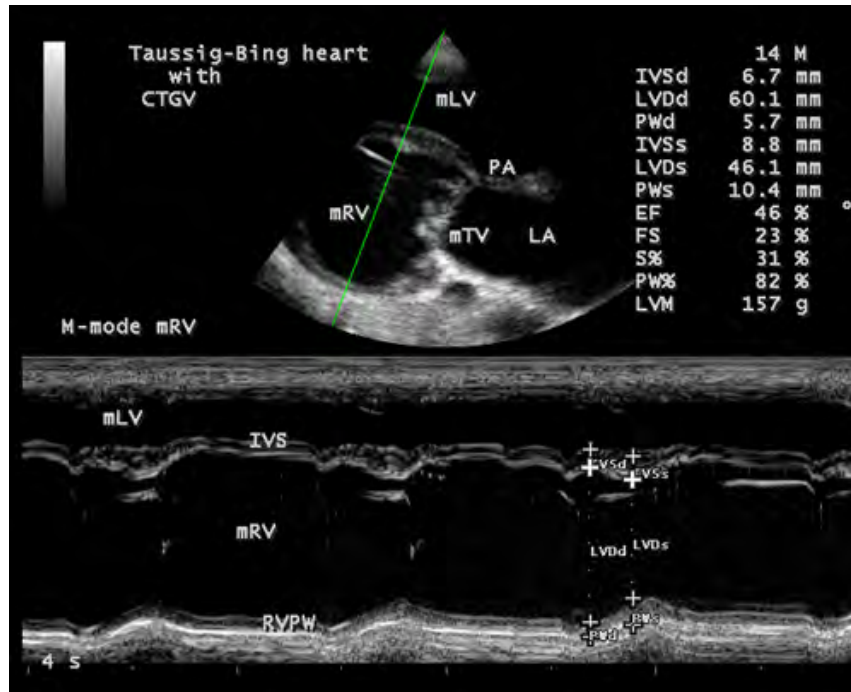
Figure 22: M-mode mRV study (green line) showing the improvement in RV function following aggressive medical treatment - EF 46%
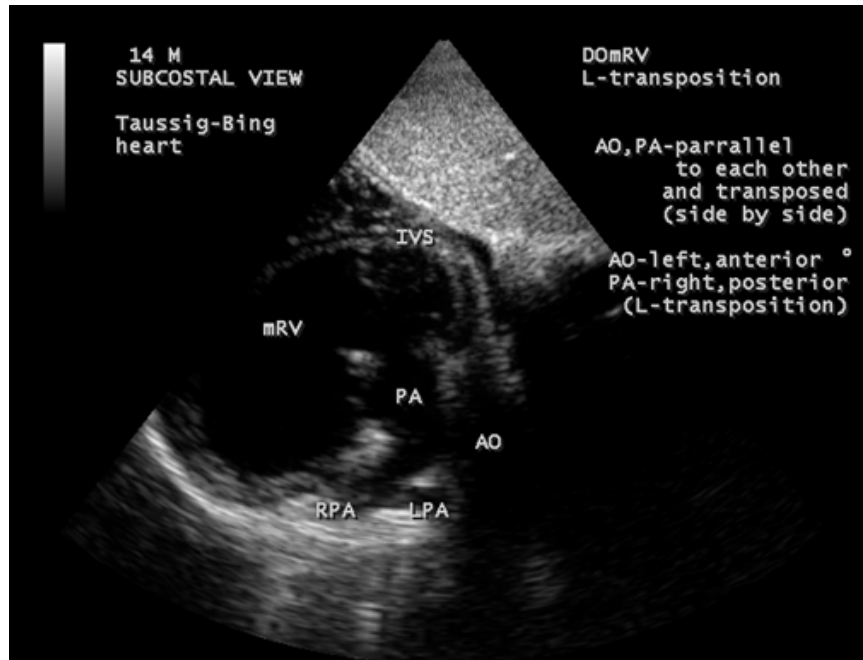
Figure 23: Subcostal view-short sweep showing the primary origin of great vessels from the morphological right ventricle (mRV) and it is side-by-side (L-transposition)
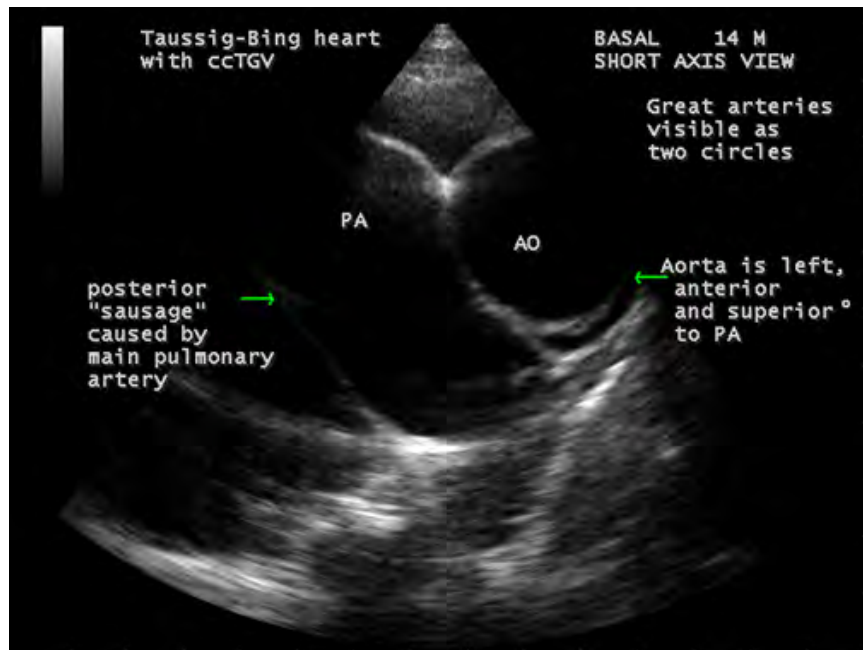
Figure 24: Basal short axis view showing the great arteries seen as circles . Right arrow-"posterior sausage" caused by main pulmonary artery. Left arrow- Aorta is left, anterior and superior to pulmonary artery (PA).
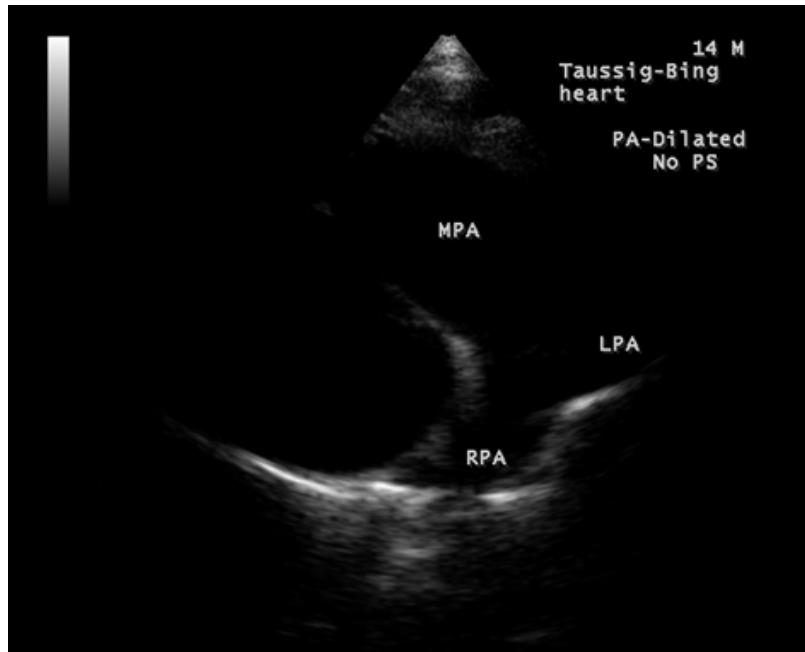
Figure 25: Showing the dilated pulmonary trunk in Taussig-Bing heart and there is no obstructive lesions. MPA- dilated main pulmonary artery.
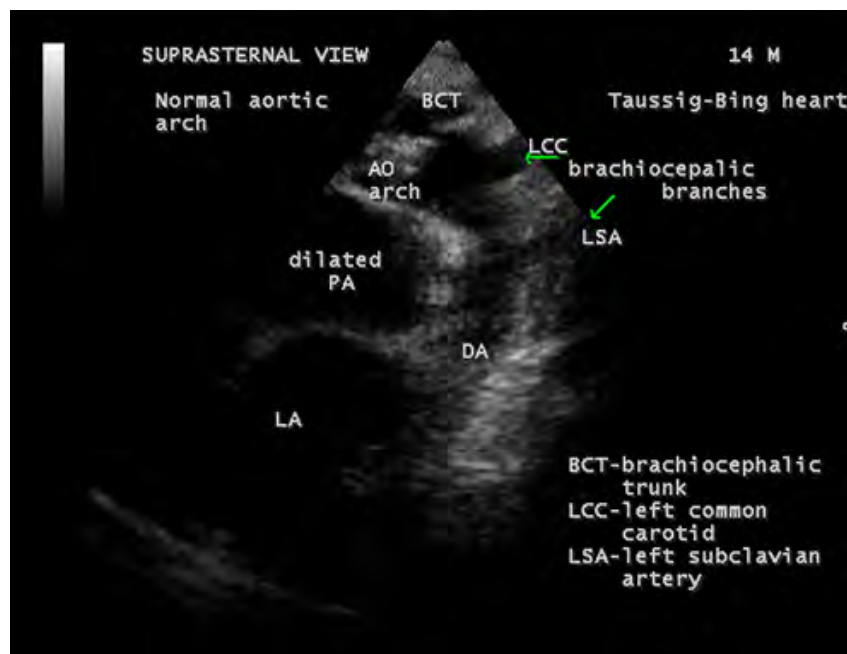
Figure 26: Suprasternal view showing the normal aortic arch. Upper arrow- left common carotid artery. Lower arrow- Left subclavian artery.

4. Discussion

4.1. Review of Literature

Taussig-Bing suspected a totally different malformation at autopsy in a five-and-a-half-year-old girl who had cyanosis since birth and died during angiocardiography after a third intravenous injection of contrast material, the diodrast, the child sat bolt upright and the heart stopped. The malformation consists of a transposed aorta, a large pulmonary artery which arises primarily from the right ventricle and partially overrides the ventricular septum, a high ventricular septal defect, and right ventricular hypertrophy. Maurice Lev was one of the first to call this anomaly as "Taussig-Bing syndrome" [8]. This anomaly was initially described in 1949 by Taussig and Bing as transposition of the great arteries. The important difference between Taussig-Bing anomaly and transposition of the great arteries were described by Richard Van Praagh in his editorial review and stressed that the absence of pulmonary-mitral continuity is the characteristic feature of this anomaly. Taussig and Bing emphasized that the overriding of pulmonary artery was an integral part of this malformation. This anomaly was eventually named as "Taussig-Bing heart" in 1950 by Lev and associates [9] and subsequently introduced the spectrum of Taussig-Bing hearts depending on the degree of pulmonary artery overriding (right-sided, intermediate, and left sided) [10]. The malalignment of infun-

dibular septum as a fundamental requisite to define these hearts, collectively termed as " Taussig-Bing complex".

4.2. Etiopathogenesis

The Taussig-Bing anomaly is relatively rare and it accounted for approximately 8% of cases in Sridaromont series [11]. In these cases, the pulmonary stenosis does not occur, the pulmonary trunk is markedly dilated as in (Figure 25) and there is a high VSD immediately subjacent to the pulmonary valve (subpulmonary) and the pulmonary valve is not in continuity with AV (atrioventricular) valve because of interposed sub pulmonary conal free wall musculature as in (Figure 14). The anatomic feature of Taussig-Bing malformation appears to result from marked and symmetrical development of both conal free walls. The semilunar valves have been protruded anteriorly to about the same extent by the underlying conal musculature and remain side-by-side, so that both great arteries arise above the anterior (right) ventricle and the failure of complete absorption of the conus or coni result in fibrous discontinuity with atrioventricular valves. During the development of the heart, the outflow tracts initially connect exclusively with the primitive right ventricle and undergo extensive remodeling to divide into aorta and pulmonary artery with connection to left and right ventricles respectively. Goor and Edwards {12} stated that double-outlet right ventricle appears to represent a primitive embryologic condition as failure to achieve conotruncal inversion and leftward shift of conus results in persistence of this condition with complete origin of both great arteries from the morphologic right ventricle. The primitive cardiac tube anchored at one end by the sinus venosus and at the other end by truncus arteriosus. The chambers of the heart develop by ballooning from the primary heart tube as atrial appendage ballooning in parallel to the atrial component of the tube and the apical component of the ventricle balloons in series from the inlet and outlet parts of the ventricular loop. The 'looping' of the straight heart tube during the 3rd week of gestation is one of the key embryologic processes leading to anatomic alignment of chambers of the heart and it normally 'loops' to the right (D-loop) resulting in normal morphological position of right ventricle to the right side. The abnormal cardiac looping to the left side (L-loop) brings the morphologic right ventricle to the left and the ventricles are reversed (ventricular inversion), the atrioventricular valves 'follow' the ventricles and the two great arteries are transposed or exit from the 'wrong' ventricle, but the systemic venous blood flows from the morphologic left ventricle to the pulmonary artery and the systemic arterial blood flows from the morphologic right ventricle to the aorta and so the circulation is corrected. Thus, during embryologic development, left-handed looping of the heart tube results in AV (atrioventricular) discordance and the aorto-pulmonary septum fails to rotate 180°, resulting in ventriculo-arterial discordance. This heart defect is called as 'corrected transposition' by Baron Rokitansky in 1875 and described as 'Rokitansky's malformation' [13] in 1957 or L-transposition (Levo-transposition) or

'ventricular inversion' and subsequently designated as 'congenitally corrected transposition'(ccTGV) by Schiebler since the correction was a gift of God, rather than the gift of the surgeon [14]. Abnormalities of the morphologic tricuspid valve (left AV valve) was seen in 90% of L-transposition in autopsy series and significant Ebstein-like malformation of left AV valve, ie, the septal leaflet is displaced inferiorly towards the cardiac apex [15] as in (Figure 7), 17 mm apical to the right-sided mitral valve (normally the distance between the tricuspid and mitral annulus is < 8 mm/m^2 (displacement index [16]), the left-sided Ebstein's anomaly [17] and it is less displacement than in the usual form of Ebstein's anomaly as in patients with concordant AV connections. The atrialization of the inlet of the morphologic right ventricle is also minimal and the "sail-like" deformity of postero-inferior leaflet of morphologic tricuspid valve is well marked in this case as in (Figure 13) which is rarely seen in left-sided Ebstein's malformation. The unusual disposition of the conduction tissue that predisposes to spontaneous AV block [18] as in (Figure 1), occurs in 1/10th of infants born with this anomaly [19],[20] with an incidence of 2%/year [21], as 45% will have developed third-degree AV block on 20-year follow up [22] and it reaches a prevalence of 10 to 15% by adolescence and 30% in adulthood [23]. In older patients, fibrotic invasion of proximal and distal bundle branches was involved in its pathogenesis. The tricuspid valve tends to remain competent during the first decade of life, but progressively become incompetent during the 2nd to 5th decade in 20 to 50% of patients and it has been implicated as a cause of ventricular dysfunction due to volume overload. Since the morphologic right ventricle is in the systemic position, it is unable to generate the necessary forces to sustain the increasing work demand, such as exercise , similar to normal systemic left ventricle [24] due to the loss of special twisting motion during systole and early diastole [25] as it lacks the helical myocyte arrangement seen in left ventricle. The RV muscle mass increase significantly since it is subjected to systemic pressure, leading to right ventricular hypertrophy (RVH) as in ECG shown in (Figure 1)

The VSD is neither membranous nor conal septal defect and it is due to its malalignment of well-developed distal conal free walls, the parietal and septal bands may separate or dissociate widely in Taussig-Bing malformation. In L-TGA, the VSDs occur in 60-80% of cases, usually perimembranous, located below the pulmonary valve, extends posteriorly and inferiorly towards the crux of the heart and opening into the inlet of the morphologic left ventricle as in (Figure 6). Endocardial cushions in the outflow tract are responsible for the formation of semilunar valves and the development of conal septum (the portion of ventricular septum between the distal ventricular outflow tracts). During conus malrotation, changes in position of anterior portion of the muscular interventricular septum and differential conus absorption leads to malposition of great artery relations. In S.L.L. type of ccTGV [26] (S-situs solitus of the viscera and atriaI, L-loop ventricle, L-transposition of great

arteries), deviation in the normal development of interventricular septum as the ventral limb of the left interventricular sulcus on the primary heart tube gives rise to a left-sided crista supraventricularis, which, determines, in part, the right ventricular morphology of the left-sided ventricle. The dorsal limb spirals towards the atrioventricular canal, giving rise to a malpositioned interventricular septum, and displaces the embryologic right ventricle to the left. The course of outlet septum establishes the alignment of VSD with great arteries. If the outflow tract is straight and parallel to the trabecular septum, the VSD becomes subpulmonary and the extreme leftward rotation of the aorta resulting the great arteries nearly in a side-by-side relationship, with the aorta to the left. The molecular and biological forces that dictate the abnormal cardiac looping in humans are not clear. Cytogenetic analysis identified the discrete chromosomal lesions in the morphogenesis of the heart and appears to be associated with subtypes of DORV. Type 1 DORV (with subaortic VSD) is associated with chromosome 22q11 deletion, the type II (with subpulmonary VSD (supracristal) - Taussig Bing type) with trisomy 13 or 18. The transcription factors such as Nkx 2.5, Gate 4 and Tbx 20 are postulated to influence the right ventricular development via IsI 1, Mef2c and Hand 2 pathways and Pitr 2, Fgf8, Fgf 10 are essential for the development of great arteries [27]. Type 1 DORV might reflect the abnormalities of neural crest cells necessary for the normal outflow tract development and the TGF$_\beta$ superfamily genes (activin type IIB receptor, noggin, TGF$_{\beta2}$), Sox4 [28], NF1, RAR genes, Endothelin converting enzyme genes (ECE-1, ECE-2) and Pax 3 genes also play a role. Mutations of Gate 4 gene has a potential for the development of type II DORV. Genes determining the body plans such as Pitx 2, CFC1, Cx43 (left-right axis determination), Hox 1.5 (rostral-caudal specification) and jefty 1, inversin (abnormalities of situs and looping) for type 111 DORV (with doubly-committed VSD) and mutations in several other genes had been implicated in few cases of human DORV pertinent to animal models [29],[30]. Recently, it is noted that there is a relationship between primary ciliary dyskinesis (PCD) and ccTGV [31]. The association between cilia and cardiogenesis was made in a baby chick and Willaredt et al [32] found a link between Sonic hedgehog signaling at the primary cilium and the role of bone morphogenic protein in the cardiogenesis of endocardial cushions. Animal studies also implicate diverse teratogens, the commonly used drugs (ephedrine, theophylline, sodium valproate, retinoic acid/vitamin A, tedral (combination of theophylline, ephedtine, phenobarbitone), other agents including bis-diamine, bromodeoxyuridine), copper citrate, environmental factors such as hair dye, air pollutants [33] and maternal diabetes are enhancing the susceptibility of DORV phenotypes.

4.3. Echocardiographic Features

Transthoracic echocardiography is the first line modality to assess the specific relations of the different segments of the heart and to determine the presence of any associated malformations. The an-

atomical designation is first assigned by demonstrating atrial position, ventricular and arterial loopings. The morphology of right ventricle is determined by the presence of course trabeculations, moderator band and the level of tricuspid valve, which is inferior (apical) to mitral valve, also give a clue to ventricular inversion as shown in (Figure 7). The morphology of tricuspid valve is characterized by a long, elongated posterior leaflet and a short septal or anterior leaflet in comparison to mitral valve, in which the posterior leaflet is short and the anterior leaflet is long and elongated as in (Figure 5). The morphologic TV (tricuspid valve) is a systemic AV valve and its septal leaflet is abnormally displaced inferiorly. This systemic AV valve (mTV) is anatomically abnormal, that predicts the occurrence of regurgitation and transthoracic echocardiography is the technique of choice to evaluate the etiology of TR (tricuspid regurgitation) and in most cases, it is so called "functional" (secondary TR (tricuspid regurgitation) due to dilatation of annulus (diastolic diameter > 40 mm or 21 mm/m^2)) as a consequence of RV dilation secondary to pulmonary hypertension. In 15 to 20% of cases, the injury to tricuspid valve can be organic and generally of rheumatic in origin. The morphologic TV (mTV) is thickened and calcified as in (Figure 7), more prominent at leaflet tips as in (Figure 11) with commissural fusion as in Figure 10, the posterior tricuspid leaflet is predominantly affected as in (Figure 12) and it is severely regurgitant as shown in (Figures 15 to 18), the annulus diameter is 30 mm (< 40 mm) as in (Figure 11). The anatomic objective such as "double discordance", ie, discordant connection at both atrioventricular and ventriculo-arterial junctions can be defined by echocardiography. The discordance between the atrium and ventricle as left atrium is connected to the morphologic Right ventricle (mRV) and the right atrium is connected to the morphologic left ventricle (mLV) was shown in (Figure 7). The origin of great arteries in relation to the septum and its spatial relationships are best visualized in subcostal coronal views. (Figure 23) shows the origin of both great arteries primarily from the morphologic RV (mRV), it is parallel to each other (normally cross each other) and the pulmonary trunk is identified by its bifurcation into right and left branches. The great arteries appear as double circles in basal short axis view as in (Figure 24) with posterior 'sausage' caused by the main pulmonary artery segment, transected tangentially and an anterior circle which is the aorta, positioned leftward, anterior and superior to the pulmonary artery [34] suggesting L-transposition. The subcostal coronal view also helps to determine the lack of fibrous continuity between the posterior semilunar valve (pulmonary) and the anterior tricuspid leaflet as in (Figures 11 and 12) and suggesting double-outlet right ventricle with a subpulmonary VSD (Taussig-Bing type). In about 50% of cases of Taussig-Bing anomaly, the congenital malformation of the aorta including coarctation, isthmic hypoplasia, interruption, patent ductus arteriosus and subaortic conus may occur [35],[36]. The aorta is identified by its brachiocephalic branches, but the aortic arch appears normal in

this patient as shown in (Figure 26).

4.4. Management

Systemic AV valve regurgitation is a potential risk factor for the development of dysfunction of the morphologic RV and early heart failure [37] and those patients without such complications (ie, in the absence of hemodynamic lesions) appear to function well into late adulthood in ccTGV [38],[39]. Medical management with diuretics, afterload reduction with angiotensin converting enzyme inhibitors such as captopril and digoxin (inotropic) is adapted for the failing systemic right ventricle [40], depending on the severity of associated malformations [41]. The role of angiotensin receptor blocker with losartan was evaluated and found to have no improvement in exercise capacity and no reduction in neurohormonal levels in patients with systemic right ventricle [42]. Beta blockers did not improve outcome in a randomized trial included pediatric patients with systemic RV dysfunction [43] and must be used cautiously in ccTGV patients as they may precipitate AV block, but an improvement in systemic ventricular function occurred after carvedilol (a vasodilatory beta blocker) administration in a 80-year old woman with ccTGV [44], Aggressive medical treatment with afterload reduction is indicated for patients with ventricular enlargement and early symptoms. The benefits of prophylactic use of vasodilators is unproven to delay or prevent systemic ventricular dysfunction.

4.5. Interventional Therapy

In patients with widened QRS complex (QRS duration > 120-140 ms, sometimes >200ms) due to bundle branch blocks, the electromechanical dyssynchrony create insufficiency in ventricular ejection and restoration of electrical synchrony with transvenous permanent para-Hisian pacing (cardiac resynchronization therapy) reduces the QRS duration and improves the hemodynamics of the failing systemic right ventricle [45],[46], but is technically challenging [47]. Complete heart block is also a common cause of death [48]. Early pacemaker placement is recommended in the setting of complete heart block if there is RV dysfunction, symptoms of bradycardia or heart failure and urgently done if it occurs during or after the surgical intervention when the bradycardia is intolerable. It can be technically challenging pertaining to the complex heart anatomy [49] such as modification in position of ventricular septum that induces a septal shift and failure of tricuspid valve coaptation [50] leading to worsening of systemic ventricular function and AV valve regurgitation.

4.6. Surgical Therapy

The evolution of surgical repair for the Taussig-Bing anomaly has progressed from atrial baffle procedures to arterial switch with VSD closure and intraventricular repair. The need for surgical interventions vary according to the associated defects and several options are available as

a. physiologic or conventional repair- which emphasizes the correcting of associated defects without addressing the discordant connections and leaves the morphologic RV to propel the systemic circulation. The tricuspid valve and morphologic RV function is the Achilles heel of the physiology of ccTGV [51],[52]. Timely performed systemic AV valve replacement may preserve ventricular function, improves long-term outcome [53] and should be done prior to significant RV dIlation in symptomatic patients and a preoperative ejection fraction of ≥ 40%. Systemic AV valve repair have had disappointing intermediate and late results [54].

b. Anatomical repair- In physiological correction, the morphological RV will likely to fail over long-term, there is an increasing trend towards anatomical correction by "switch procedures" introduced in 1987 by Ilbawi and colleagues with an aim to utilize the morphologic LV as a systemic pumping chamber and the mitral valve as the systemic valve [55]. The goal of anatomic correction is the rerouting of the pulmonary venous return to mLV(morphologic LV) and aorta, the systemic venous return to mRV(morphologic RV) and pulmonary artery that achieving a normal anatomical pattern of circulation., ie, morphologic LV to aorta and morphologic RV to pulmonary artery.

It represents a group of procedures in which AV discordance is corrected by an inflow, baffling procedure at atrial level, called as atrial or venous switch (Mustard or Senning) and the ventriculo--arterial discordance is corrected by either an outflow procedure at arterial or ventricular level, called as arterial or ventricular switch or by translocation of the aortic root (Jatane procedure) and collectively termed as " double-switch". The choice of procedures depending on the underlying anatomy of LVOT/or morphology of VSD [56]. A modification of atrial switch procedures to a hemi-Mustard technique can be performed to overcome the high early morbidity and mortality in more complex ccTGV patients to decrease the surgical challenge [57]. The indication for anatomic repair for those with a "balanced" systemic and pulmonary blood flow without clinically significant cyanosis is still unclear. Current indication for 'double switch' operation include patients who have developed symptoms attributed to tricuspid regurgitation or RV dysfunction or both [58]. The anatomic repair is currently the first choice for most surgeons in treating ccTGV [59], only meant for patients with a favourable anatomy [60] and it remains the best choice for TGA type of DORV [61]. Early prophylactic pulmonary banding [62] as a bridge to 'double switch surgery' related to LV retraining in selected asymptomatic young infants with isolated ccTGA is a challenging practice to expand anatomic repair in those patients who suffer from persistent late LV failure due to LV retraining.

For patients with Taussig-Bing type of DORV, the 'arterial switch' operation still appears to be the procedure of choice and can be performed in the neonatal period [63] and also in patients with all types of great artery anatomy without ventriculotomy. The arterial switch for Taussig-Bing heart was first reported in 1981 [64]. The

issue of great artery anatomy was first addressed by Yacoub and Radley- Smith [65] and many surgeons have thought that the side-by-side great arteries are most suitable for intraventricular repair (Kawashima repair) [66],[67], which is also applicable to unusual coronary artery anatomy or a pulmonary valve that is not considered adequate to function as a systemic valve. The antero-posterior great arteries are most suitable for arterial switch with a closure of VSD.. The distance between the tricuspid and pulmonary valve for an intraventricular tunnel [68] is more important than the great artery relations and in side-by-side great arteries, the conduit can pass posteriorly and allow unimpeded flow to the pulmonary valve, ie, by tunneling the left ventricular flow posterior to the pulmonary valve. The operative mortality for arterial switch is 13%, 3% for intraventricular repair and late mortality for both series is 5 to 6%.

a. Single - ventricle palliation - such as 'Fontain circulation'(redirection of systemic (deoxygenated) blood into the pulmonary artery without traversing a ventricle) should be considered in more complex and unfavourable anatomy due to small RV, chordae straddling and remote VSD, however, it may affects the organs outside the heart leading to cirrhosis, protein loosing enteropathy and plastic bronchitis [69]

4.7. Outcome

In patients without tricuspid valve regurgitation, the survival rate at 20 years is > 90% and it is < 50% in regurgitant lesions. Unfortunately, the patients are often referred late for surgery when a progressive RV dysfunction is already initiated, however, if the surgery is performed when the RV ejection fraction is > 40%, the patients do reasonably well. In addition, a number of reports of ccTGA in adults come to medical attention for either cardiac or non cardiac reasons in 6th to 8th decade, indeed, the right ventricle can function as a systemic ventricle for many tears [70].

5. Case Analysis

The 14-year old boy was presented with features of "double discordance" (Figure 7) with primary origin of L-transposed great arteries from the left-sided morphologic right ventricle (Figure 23) and a pulmonary-tricuspid discontinuity with well developed pulmonary conus (Figure 14) and a malaligned subpulmonary VSD (Figure 10), suggesting a "Taussig-Bing heart" with L-transposition. The visceral situs was normal (Figure 4) and there Is a marked RV dilatation (Figure 11) with severe tricuspid regurgitation (Figures 15 to 18) due to rheumatic etiology (Figures 8 to 10) and so it is designated as "Rheumatic Taussig-Bing heart". The septal malalignment in situ solitus alters the cardiac hemodynamics and conduction pathways, leading to increased risk of complications. The atrial and ventricular septa are malaligned and the AV

node cannot penetrate the ventricular mass, indeed, a secondary node penetrates the anterior septum, predisposing to high grade atrioventricular block [71] (Figure 1) with a tolerable heart rate (50 bpm) due to congenital etiology. The septal malalignment in situs solitus also leads to a high prevalence of Ebstein-like anomalies and 50% of ccTGV patients had abnormal tricuspid valve , compared to normal in situs inversus group. The left-sided Ebstenoid-like malformation of mTV as in (Figure 7) (apical four chamber view) is due to septal malalignment, mimicking as apical displacement of septal tricuspid leaflet which is normally attached to annulus as in Figures 8 and 9 shown in subcostal views with a subpulmonary VSD (Figure 10) as the result of this septal malalignment. On aggressive medical management with anti-failure measures and penicillin prophylaxis, the severe tricuspid regurgitation decreases as shown in (Figures 19) and 20. It is a primary tricuspid regurgitation due to rheumatic involvement of mTV and the tricuspid annulus is not dilated (< 40 mm) as in (Figure 11). The mRV function improves from the severe dysfunction (Figure 21- EF 36%) to a moderate degree (Figure 22- EF 46%). The boy remains acyanotic with an improvement in hemodynamic status, the medical measures are continued with lifelong penicillin prophylaxis and advised periodic echocardiographic evaluation to assess the ventricular function. Since the left-sided morphologic RV is a systemic pumping chamber, it is hypertrophied (Figure 1) and may deteriorate in function on long-term follow-up. Since the both ventricles are adequate without any outflow tract obstructions (Figures 25, 26) and the great arteries are in side-by-side, the Kawashima intraventricular repair is the preferred surgical option with tricuspid valve replacement in this boy. The pacemaker implantation is necessary when the heart rate becomes intolerable especially during surgical procedures.

6. Conclusion

Taussig-Bing heart is one of the variants of double-outlet right ventricle (DORV) [72] and it constitues < 10% of cases of DORV. It is considered as a spectrum of anomalies, unified by a juxta-pulmonary VSD with malalignment of infundibular septum as double outlet right ventricle (Type II DORV- Taussig-Bing type) at one end and a discordant ventriculo-arterial connection at the other end. The options of surgical correction are determined by internal morphology of the lesions. The arterial switch operation appears to be preferred [73] and in selected cases, excellent early and intermediate survival had been reported with Kawashima intraventricular repair [74] recently. Vitamin E administration results a notable reduction in the severity of cardiac malformations in animal models , especially in diabetic pregnant mice due to its antioxidant effect [75].

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