Surgical Management of Lesions encountered in the Setting of the Retroaortic Left Brachiocephalic Vein

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Abstract

The present perspective is a synthesis of 80 published investigations in the setting of a retroaortic left brachiocephalic vein, described in 250 patients. Clinical presentation, radiographic findings, ultrasonographic findings, saline contrast echocardiography, computed-tomographic angiocardiography, magnetic resonance imaging, and angiocardiography provided the diagnostic information used to define the disease entity prior to considering the surgical approach to the associated cardiac anomalies. We have also addressed several issues concerning the influence of isomerism, the establishment of diagnosis, and its importance in various surgical and interventional procedures. Although the retroaortic left brachiocephalic vein is asymptomatic, its recognition during clinical investigation should raise the possibility of an association with other malformations, especially right aortic arch, ventricular septal defect, and anomalies of the outflow tracts. We submit that an increased appreciation of this venous anomaly may facilitate surgical planning, endovascular procedures, placement of central venous lines, and transvenous pacemakers.

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Running Title: Retroaortic left brachiocephalic vein

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Abstract

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Although the retroaortic left brachiocephalic vein is asymptomatic, its recognition during clinical investigation should raise the possibility of an association with other malformations, especially right aortic arch, ventricular septal defect, and anomalies of the outflow tracts. We submit that an increased appreciation of this venous anomaly may facilitate surgical planning, endovascular procedures, placement of central venous lines, and transvenous pacemakers.

Key words : Aberrant left brachiocephalic vein, Aortic arch anomalies, Conotruncal cardiac anomalies, Double aortic arch, Retroaortic anomalous left brachiocephalic vein, Right aortic arch, Tetralogy of Fallot

Introduction

The left brachiocephalic vein usually courses obliquely downward, passing anterior to the aortic arch and its own brachiocephalic branches. It joins the right brachiocephalic vein to form superior caval vein. Rarely, the left brachiocephalic vein takes an anomalous course. It extends lateral to the aortic arch, enters the gap between the extrapericardial aorta and the right pulmonary artery, and crosses the mediastinum posterior to the ascending aorta and anterior to trachea. It then joins the superior caval vein between the azygous vein and superior cavoatrial junction.[1-5] This pattern is usually described as the retroaortic left brachiocephalic vein (Figures 1A-1F).[1-6]

It is a rare anomaly, first described by Kerschner in 1888.[7] The first angiographic description was provided by Roberts and associates in 1951, with Cloez and associates recognizing it echocardiographically in 1982. Webb

and associates described computed tomographic findings, again in 1982, while Fujimoto and associates showed magnetic resonance findings in 1992.[1,8-10] In 1992, Takada and associates described a double circumaortic left brachiocephalic vein.[11]

As far as we are aware, around 250 cases have now been reported since 1980.[1-80] When found in isolation, the anomalous course is of no clinical importance. Recognition of in the setting of associated lesions, in contrast, is important. This is because it can be mistaken for a persistent left superior caval vein, or a vertical vein in the setting of totally anomalous pulmonary venous connection. In this review, we evaluated all clinical, as well as necropsy, studies describing this venous anomaly with and without coexisting congenital heart diseases. We have emphasized its potential implications for the cardiac surgeon.

Methods

We searched the literature to identify the described instances of retrotracheal/ retroaortic/anomalous/aberrant left brachiocephalic vein, along with circumaortic double left brachiocephalic vein. We evaluated all clinical as well as autopsy studies, seeking to establish accounts of the anatomy, the prevalence of isomerism, problems in establishment of diagnosis, and the variations in the associated cardiac and aortic arch anomalies. We then assessed its influence on surgical techniques used in treatment, and their outcomes. The search engines employed were PubMed, Google Scholar, Cochrane Database for Systematic Reviews, Cochrane Central Register of Controlled Trials, Ovid Medline, ACP Journal Club, Ovid EMBASE, and Database of Abstracts of Review of Effectiveness in all languages.

This strategy provided 80 investigations that provided the best answers to these topics. We have then synthesized all available data, establishing issues of concern, diagnostic clues, and various techniques performed to repair the primary as well as concomitant anomalies, hoping to improve future surgical management.

Due to limited sample sizes, heterogeneity of clinical state at the time of surgical intervention, and difficulties in selection of appropriate cardiac quantifiable end points, we were unable to achieve a meta-analysis.

Results

Incidence

Due to its rarity, the true incidence with either usual or mirror-imaged atrial arrangement, or in association with isomerism of the atrial appendages, is difficult to establish. The incidence in patients with congenital heart diseases was reported as 0.2% in necropsy studies,[7,18,23,37-42,63-66] 0.5%-0.98% in echocardiographic series,[11,21] and 1.7% when using computed tomography.[4,19,41,71] Its occurrence without any associated cardiac or aortic arch anomaly is extremely rare. Its prevalence is estimated at 0.02%.[14-17,33]

Demographics

The age of reported patients at their initial diagnosis, and at necropsy, ranged from 1 day to 84 years, with a median of 71.5 months, and an interquartile range from 14 to 498 months. The great majority were below 18 years of age.[1-80] The male to female ratio in patients with congenital heart diseases was equal, but males predominated by 1.7 to 1 in reported isolated cases. There was no identifiable regional or ethnic predominance. Patients with isomerism of either the right or left atrial appendages had an increased prevalence of the anomaly, at around 13%.[4,11-14] As emphasized, it is exceedingly rare to find the lesion in the absence of congenital heart diseases or aortic arch anomalies.[14-17]

Diagnosis

Diagnosis of the isolated lesion is difficult, since it does not produce any symptoms.[23,62,63,66] Presentation, therefore, is usually based on the discovery of associated cardiac anomalies. In earlier reports, the diagnosis was most often established at necropsy.[7,18,23,37-42,63-66]

The chest roentgenogram findings vary according to the associated anomalies. Association of a curved border in the superior mediastinum on frontal chest radiographs, or a high aortic arch on the lateral projections, is suggestive, although not diagnostic.[3]Diagnosis can usually be made by transthoracic ultrasonography,

and is enhanced by saline contrast transthoracic or transesophageal echocardiography.[11-13,18,19,21-25] The ultrasound "window" may be of poor quality in cases of thymic hypoplasia or agenesis.[14] Visualization of the frontal section of the aortic arch allows recognition of the entire course of left brachiocephalic vein. If the lesion is suspected, then the right anterior oblique view is used for a left aortic arch, and the left anterior oblique view for a right aortic arch.[12]

Using suprasternal views, contrast echocardiography with injection in left arm vein usually demonstrates the course with greater accuracy. Doppler recording allows avoidance of confusion with other structures, particularly the right pulmonary artery or the unusual atrial appendages as found in left isomerism (Figures 2A and 2B).

Three-dimensional computed tomographic angiocardiography, and magnetic resonance imaging, are helpful in ascertaining the diagnosis, and in demonstrating the relationship of the vein to the adjacent structures (Figures 1A-1F, 3A-3C, 4A-4C).[3-5,9,10,14,15,17,22,25,27-29,32-36,43-46,59,69]Cardiac catheterization and angiography may still be indicated in doubtful cases, and for evaluation of associated cardiac anomalies.[13,20,28,48,55,57,74] Computed tomography and magnetic resonance imaging similarly come into their own for the assessment of associated lesions.

Surgical anatomy

In most instances, it courses from the left, extending posterior to the aortic arch, and traversing from left-to-right anterior to the trachea, and antero-superior to the bifurcation of the pulmonary trunk, before crossing posterior to the ascending aorta to terminate in the superior caval vein (Figures 1A-1F, 2A-2B). Characteristic features have been identified in that, first, the superior caval vein is shorter than the normal case, and second, the azygous vein does not drain into the right brachiocephalic vein.[37] Very rarely, the retroaortic vein can join the azygous vein before entering the superior caval vein.[31] In two instances, the vein has been found to drain directly into the left atrium, with production of severe cyanosis.[12,13]

We encountered 6 descriptions of the duplication of the vein, the branches encircling the aorta, and one report of persistent left superior caval vein with duplication of the left brachiocephalic vein.[17,31,43,45-47] In one of the cases, the anterior and thinner vein coursed above the aortic arch, anterior to left common carotid artery, and then passed between the left common carotid artery and brachiocephalic trunk to join the right superior caval vein. The posterior thicker retroaortic vein coursed posterior to trachea and esophagus and joined the azygous vein at the level of the second thoracic vertebra before draining to the right superior caval vein.[31]

Some investigators have recognized three variations in course relative to the arterial duct or its ligamentous remnant, with the vein either passing anteriorly or posteriorly, or passing behind one or more of the main aortic branches but retaining a normal relationship to the aortic arch .[18,21] Another group, however, having assessed several series, argued in favour of four patients.[17,19,38-42] They suggested that the vein could cross the midline above the aortic arch, but posterior to the origin of great arteries (Figures 5A and 5B). In their second pattern, the vein passes beneath the aortic arch, but above the bifurcation of the pulmonary trunk, passing in front of the arterial duct (Figure 5C). The third pattern was characterized by a course posterior to the arterial duct, with crossing the midline beneath the aortic arch and above the pulmonary arteries (Figure 5D). In the final pattern, the vein crosses the midline behind the pulmonary trunk away from the arterial duct (Figure 5E). It subsequently became possible to identify two more patterns. In the first of these, the vein takes a more posterior route relative to the trachea and esophagus, subsequently joining the azygous vein before draining to superior caval vein (Figure 5F).[31] The sixth pattern is produced by duplication of the vein (Figure 5G).[31,43] There is then additional variation in the setting of duplication. The anterior vein has been described as passing above the aortic arch and coursing anterior to the left common carotid artery and brachiocephalic arteries, before draining into the superior caval vein. The posterior vein runs below the aortic arch and courses posterior to the descending aorta. Both veins in this variant drain separately into the superior caval vein (Figure 5H).[17,18,23,43-47]

From the 250 reported cases we were able to identify, three-quarters with associated congenitally malformed

hearts. Of these 189 patients, all but 7 had usual atrial arrangement. Right isomerism was reported in 5 patients, with two patients having left isomerism.[4,12,13,23] Detailed description of the cardiac anomalies was provided for 145 patients.[48,49]Of these, almost two-thirds had tetralogy of Fallot, or its variants, with just over one-sixth having ventricular septal defects with pulmonary atresia.[2,5,6,11,14,22,23,27-29,49-52] Over four-fifths had malformations involving the outflow tracts. Among these patients, two-thirds had a right aortic arch, with two patients having a cervical aortic arch, and one patient a double aortic arch.[14,22,27-29,50-53] In those without congenital heart diseases, one-sixth had right aortic arch[3,4,27], just under one-tenth had double aortic arches,[27,28,53]and two-fifths had cervical aortic arches.[22,52,54,55]

Associated cardiac anomalies

Other reported associated cardiac anomalies include tetralogy of Fallot with coarctation of aorta, unroofed coronary sinus, a persistent left superior caval vein, double outlet right ventricle, common arterial trunk, ventricular septal defect, interrupted aortic arch, tricuspid atresia, totally anomalous pulmonary venous connection, ventricular septal defect with coarctation of aorta, pseudocoarctation of aorta, aortopulmonary window, hypoplastic left heart syndrome, right superior pulmonary vein to superior caval vein, anomalous subclavian artery from patent arterial duct, superior-inferior ventricles, transposition, pseudocoarctation of aorta, origin of left vertebral artery from arch of aorta, right isomerism, left isomerism and distal aortic arch aneurysm.[2,5,11-13,19,22,23,25,39,49,55-59,61-71] The deletion of chromosome 22q11.2 was confirmed in five reported patients, while three patients had DiGeorge syndrome. The vein has also been reported as draining into a left superior caval vein with partial unroofing of the coronary sinus, and co-existing with a left superior caval vein in the setting of a normal coronary sinus.[13,79]

Surgical approach and management

Although the anomalous course of the vein, in itself, does not mandate surgical intervention, its presence can affect the surgical technique when dealing with the associated cardiac anomalies. Its presence may complicate exposure of pulmonary arteries, for example, when creating a systemic-to-pulmonary arterial shunt.

Because of its course posterior to ascending aorta or its branches, and its proximity to adjacent trachea and esophagus, the surgical view may be limited. Moderately hypothermic cardiopulmonary bypass at 32°C with cold blood cardioplegia has been the most popular technique used by several investigators, depending on the associated cardiac anomalies. Deep hypothermic circulatory arrest has been employed for separate cannulation of the caval veins, or exposure of the aortic arch or pulmonary arteries. This technique has also been suggested to be impractical because of the short superior caval vein, and the small operating field.[5,56,75] The principles of surgery are dictated by the concomitant cardiac anomalies. Multiple techniques have been reported.

Association with coarctation of aorta

This involved anterior translocation of the brachiocephalic vein. Under deep hypothermic circulatory arrest at 25°C, the arterial duct was ligated, the aortic arch was transected, and the anomalous vein was translocated in front of the aortic arch. The hypoplastic aorta was ligated beyond left subclavian artery, and ductal tissue was resected. The continuity of ascending aorta was re-established by a "semilunar" anastomosis, anastomosing the descending and ascending aorta.[5]

Direct anastomosis of interrupted aortic arch between the left common carotid and subclavian arteries

In this report,[59] an associated ventricular septal defect was closed using a polytetrafluoroethylene patch. The presence of the retroaortic vein did not impair the performance of direct anastomosis.

Total aortic arch replacement

Using antegrade selective cerebral perfusion and circulatory arrest at 20°C, the vein was preserved by complete exposure and retraction.[32]

Transection at the cavoatrial junction and left superior cavopulmonary anastomosis

This procedure was described in a 2 year old child with tricuspid atresia. Under circulatory arrest at 18°C, the anomalous vein was transected at its junction with superior caval vein. The distal end was oversewn. The proximal end was mobilized and was anastomosed to left pulmonary artery. The azygous vein was ligated. The pulmonary trunk was divided at the level of previous banding. A hemi-Fontan type of right cavopulmonary anastomosis was then performed, along with a Damus-Kaye-Stansel procedure. A year later, the child underwent a fenestrated lateral tunnel Fontan procedure.[56]

Extensive pulmonary arterial reconstruction using the vein

This innovative procedure was again used in a patient with tricuspid atresia, along with pulmonary atresia and stenosis of confluent pulmonary arteries. The parallel course of the anomalous vein facilitated pulmonary arterial reconstruction. The ascending aorta was transected under circulatory arrest at 18°C. A side-to-side anastomosis of the vein and pulmonary arteries was formed, along with right bidirectional Glenn (Figure 6).[75]

Bilateral bidirectional Glenn without cardiopulmonary bypass

This procedure was again used in the setting of tricuspid atresia. Since the superior caval vein was very short, and the retroaortic left brachiocephalic vein was located behind the ascending aorta, the superior caval vein was cannulated into the right brachiocephalic vein. Because of the retroaortic location, both the caval veins required extensive mobilization. A bilateral bidirectional Glenn was performed using a temporary stent between the superior caval vein and the right atrium.[48]

Retroaortic brachiocephalic vein with tetralogy of Fallot and pulmonary stenosis/pulmonary atresia

The great majority of the reported cases found in association with tetralogy of Fallot with pulmonary stenosis or pulmonary atresia have required extensive pulmonary arterioplasty, and insertion of a transannular patch. Systemic-to-pulmonary artery anastomosis using a polytetrafluoroethylene graft has also been performed in case of borderline pulmonary arteries.[2,5,6,11,14,22,23,49-52]

Rechanneling of supracardiac totally anomalous pulmonary venous connection

The anomalous pulmonary venous connection drained through an ascending vertical vein, which ultimately joined the retroaortic brachiocephalic vein. In this case, surgical ligation of the brachiocephalic vein could have proved disastrous.[23]

Biatrial drainage with persistent left superior caval vein

In this case, the retroaortic brachiocephalic vein drained to both the left atrium and the superior cavoatrial junction, along with a persistent left superior caval vein. A 5 cm long window like communication between the left superior caval vein and the left anterior appendage was repaired by direct suture.[13]

Surgical Results

The overall early mortality for patients undergoing repair of congenital heart diseases was 3.4%. The causes of mortality in the early postoperative period were congenital heart diseases in two patients, and cerebrovascular accident in one patient. The modalities used after establishing diagnosis were not clearly described in two-thirds of the patients. These investigations mostly dealt with multimodality imaging studies and necropsy data.[7,18,23,39,40,63,64,66] There is documentation of the anomalous vein in 22 patients dying of diverse causes, namely congenital heart diseases, cerebrovascular accidents and malignancies (Table E1).[7,18,37,39-43,64,66]

Discussion

The anomalous retroaortic left brachiocephalic vein is a rare congenital systemic venous anomaly. It has both clinical and surgical fascination. That we were able to identify only 250 cases with this disease since 1980 underscores its rarity, but highlights the need for a high index of suspicion of its existence.[37,39,40-42,64-66]

Normally, the left brachiocephalic vein enters the superior caval vein above the insertion of azygous vein and the connection is between left and right anterior cardinal components. When taking a retroaortic course, the

vein enters the right superior caval vein below and behind the aortic arch, and below the entrance of azygous vein.[18] The anomalous vein, therefore, is embryologically distinct from the normal left brachiocephalic vein, as well as having a different spatial pathway and anatomical relations. The question of terminology, therefore, could be debated at length. Because the ultimate connections at either end are identical, it seems reasonable to refer to the structure as an anomalous brachiocephalic vein.

Although of interest anatomically, the retroaortic position is physiologically inconsequential. With echocardiography, it is most easily seen using suprasternal long-axis projections (Figures 2A and 2B). Radiologically, the anomaly can be documented when catheters are placed in both brachiocephalic vein and aorta or with left superior venography. Detection is obviously possible by computed tomographic-angiography, or magnetic resonance imaging (Figures 1A-1F, 3A-3C). On non-contrast-enhanced computed tomographic scans, the subaortic portion of anomalous left brachiocephalic veins mimics enlarged lymphnodes.[72,73]

Regardless of its pathogenesis, recognition of this anomaly is important in various clinical settings. The preoperative recognition during clinical investigation raises the possibility of associated congenital cardiac malformations, especially right ventricular outflow tract obstruction, tetralogy of Fallot, with pulmonary stenosis or pulmonary atresia, right aortic arch, ventricular septal defect and totally anomalous pulmonary venous connection. Conversely, when the diagnosis of tetralogy of Fallot with pulmonary stenosis or pulmonary atresia is made, the anomalous position of left brachiocephalic vein should be looked for, especially in patients with a right aortic arch.

To the radiologist, and echocardiographer, the descending portion of an anomalous brachiocephalic vein must be differentiated from a persistent left superior caval vein or an ascending vertical vein in supracardiac totally anomalous pulmonary venous connection, and a left partially anomalous pulmonary venous connection.[3,17,18,23]

Its middle portion needs to be differentiated from the bifurcation of the pulmonary trunk.[3,18,23] The retroaortic crossing segment of the anomalous vein may be misinterpreted on unenhanced computed tomographic scan as enlarged lymphnodes, an elevated right pulmonary artery in patients with hypoplastic or atretic central pulmonary arteries, or an early branching upper lobe pulmonary artery on cross-sectional echocardiography.[4,11,17-19,21,26]

Carefully tracing this vascular channel through sequential images is the key to differentiation. Computed tomography, along with magnetic resonance imaging, are 100% sensitive in diagnosing the anomaly.[72,73]

During surgery, knowledge of the low entry site of the brachiocephalic vein might prevent is inadvertent obstruction when direct cannulation of the superior caval vein is necessary.[12] Separate cannulation of the left superior caval vein is not necessary. Because of the anomalous entry of the retroaortic brachiocephalic vein at the superior cavoatrial junction, the use of direct superior caval venous cannulation must be done with extreme precaution to avoid cannula-induced venous obstruction, or damage to arterial supply of the sinus node.[2]

In a case with tricuspid atresia and hypoplastic pulmonary arteries, the vein was used to augment the pulmonary arteries during cavopulmonary anastomosis.[74] The other situations where this anomaly might affect surgical technique is during superior cavopulmonary anastomosis for staged Fontan procedure. Its presence may also complicate exposure of the pulmonary arteries.[10,18]Its presence may also complicate the exposure and ligation of the persistently patent arterial duct, coarctation of aorta and distal aortic arch aneurysm.[2,5,17,18,50] While performing a systemic-to-pulmonary arterial anastomosis, its presence might complicate exposure of the pulmonary arteries.[2] It appears to be associated with a greater degree of obstruction in right ventricular outflow tract in the setting of tetralogy of Fallot. Its presence, therefore, should alert the surgeon to the possible requirement for a more extensive transannular parch to accomplish a satisfactory repair.

Conclusion

The retroaortic left brachiocephalic vein is a rare congenital systemic venous anomaly, usually associated with malformations involving the outflow tracts. It is also strongly associated with aortic arch anomalies,

such as right aortic arch and cervical aortic arch, patent arterial duct, coarctation of aorta, and distal aortic arch aneurysms. Although it does not cause clinical symptoms, knowledge of its presence is important when surgical interventions are required in patients with congenitally malformed hearts, especially in the setting of tetralogy of Fallot, a functionally univentricular heart, right aortic arch, and other aortic arch anomalies.

Author's contribution

Author's name	Concept/ design	Data analysis/ interpretation	Drafting article	Critical revisi
Ujjwal Kumar Chowdhury	?	?	?	?
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Niwin George	?	?	?	?
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81.

Figure Legends

Figures 1A-1F : Axial computed-tomographic angiography images (A to C, cranial to caudal) shows the left brachiocephalic vein (L) crossing posterior to the aorta (A) and joining the right brachiocephalic vein (R) to form the right-sided superior vena cava (RSVC). Volume rendered images (D, anterior view; E, anterior view with aorta digitally subtracted and F, posterior view with aorta digitally subtracted) demonstrates the retroaortic course of the left brachiocephalic vein.

Figures 2A, 2B : Transthoracic Echocardiogram images with color Doppler interrogation demonstrating the retro-aortic innominate vein. Panel A shows parasternal short axis view at the level of ascending aorta (AO). The brachiocephalic vein (IV) is seen coursing posterior to the ascending aorta and anterior to right pulmonary artery. Panel B shows the suprasternal view in which the brachiocephalic vein (IV) is seen coursing below the arch of aorta. The right pulmonary artery (RPA) is visualized coursing posterior and inferior to the retro-aortic brachiocephalic vein.

Figures 3A-3C : Four chamber image (A) and oblique sagittal image (B) shows presence of subaortic ventricular septal defect (*) with aortic (A) override in a patient of tetralogy of Fallot. Right ventricular (RV) hypertrophy is also noted. Oblique coronal image (C) shows presence of severe infundibular stenosis (arrowhead).

Figures 4A-4C : Volume rendered images (A to C) show presence of subaortic ventricular septal defect (*) with aortic (A) override. Right ventricular (RV) hypertrophy is noted with severe infundibular stenosis (arrowhead).

Figure 5A-5H : Diagram illustrating the normal and abnormal course of left brachiocephalic vein

A: Normal course of left brachiocephalic vein in front of the great arteries.

B: The anomalous left brachiocephalic vein crosses the midline above the aortic arch posterior to the origin of the great arteries.

C: The anomalous brachiocephalic vein passes beneath the aortic arch over the pulmonary artery bifurcation in front of the arterial duct.

D: The anomalous brachiocephalic vein passes posterior to the arterial duct, beneath the aortic arch and above the pulmonary artery.

E: The anomalous brachiocephalic vein crosses behind the main pulmonary trunk away from the arterial duct.

F: The anomalous brachiocephalic vein passes posterior to trachea and esophagus and joins the azygous vein.

G: The anomalous brachiocephalic vein divides into two branches. The anterior branch is located above aortic arch, behind the brachiocephalic trunk. The posterior branch crosses posterior to esophagus and joins the azygous vein before draining to right superior caval vein.

H: The circumaortic anomalous brachiocephalic vein divides into two branches. The anterior branch courses above aortic arch anterior to great arteries. The posterior branch courses below the aortic arch and posterior to descending thoracic aorta. Both branches joined the right superior caval vein separately.

Figure 6 : Diagrammatic representation of use of retroaortic left brachiocephalic vein in pulmonary artery reconstruction by creating a wide side-to-side anastomosis. The child had tricuspid atresia, pulmonary atresia with a left modified Blalock-Taussig shunt, a stented arterial duct and blocked central aortopulmonary shunt to the right pulmonary artery.

Abbreviations: ACV: Anterior cardinal vein, AO: Ascending aorta, APS: Aortopulmonary shunt, AV: Azygous vein, BCA: Brachiocephalic artery, CCV: Common cardinal vein, DTA: Descending thoracic aorta, ITCP: Inferior transverse capillary plexus, IV: Brachiocephalic vein, LBCV: Left brachiocephalic vein, LCCA: Left common carotid artery, LIJV: Left internal jugular vein, LOM: Ligament of Marshall, LPA: Left pulmonary artery, LSA: Left subclavian artery, LSV: Left subclavian vein, LV: Left ventricle, P: Pulmonary trunk, PA: Pulmonary artery, PAD: Patent arterial duct, PCV: Posterior cardinal vein, RA: Right atrium, RPA: Right pulmonary artery, RSVC: Right-sided superior vena cava, RVCV: Right brachiocephalic vein, STCP: Superior transverse capillary plexus, SV: Sinus venosus, SVC: Superior caval vein, VOM: Vein of Marshall,

Table E1: Summary of the published investigations documenting the diagnosis of retroaortic anomalous brachiocephalic vein and its management (clinical and necropsy studies)

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
1.	Gerlis LM and Ho SY et al[18]	1989	5 years 11 months/F	Case 1 (Necropsy): Died of cerebral ischemia-postop TOF. Postmortem-TOF, several large MAPCAs, RAA, retroaortic BCV joining RSVC below azygous vein above the right superior cavoatrial junction	Postoperative TOF	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			Male infant	Case 2 (Necropsy): Died of complex cardiac malformation. PM examination: concordant AV connection, DORV, VSD, destroposed aorta, PAD, retroaortic BCV anterior to the arterial duct between the arch and pulmonary trunk, joined RSVC between azygous vein and right atrium	Not applicable	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			7 week/F	Case 3 (Necropsy): Died attempted surgical correction-common arterial trunk. Concordant AV connection, subtruncal VSD, quadricuspid truncal valve, retroaortic BCV below the aortic arch, joined below the azygous vein	Not applicable	Not applicable	Not applicable
2.	Kerschner L et al[7]	1988	? child, ? sex	Necropsy, relation to arterial duct ?, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable
3.	Daser P et al[39]	1902	68 years/M	Necropsy, anterior to arterial duct, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
4.	Ghon A et al[64]	1908	$4\frac{1}{2}$ months/F	Necropsy, posterior to arterial duct, right aortic arch, absent 4th aortic arch, isolated LSA	Not applicable	Not applicable	Not applicable
5.	Nutzel H et al[40]	1914	74 years/M	Necropsy, left aortic arch, right superior pulmonary vein to RSVC	Not applicable	Not applicable	Not applicable
6.	Martin CP et al[65]	1931	17 years/M	Necropsy, right aortic arch, anterior to arterial duct, TOF	Not applicable	Not applicable	Not applicable
7.	Walter L et al[41]	1931	19 years/M	Necropsy, anterior to arterial duct, left aortic arch, subthyroid venous anastomosis	Not applicable	Not applicable	Not applicable
8.	Adachi B et al-[37]	1933	41 years/M	Necropsy, posterior to arterial duct, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
9.	Adachi B et al[37]	1933	20 years/M	Necropsy, anterior to arterial duct, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable
10.	Friedman SM et al[42]	1945	66 years/M	Necropsy, anterior to arterial duct, left aortic arch, left jugular vein anomaly	Not applicable	Not applicable	Not applicable
11.	Roberts JR et al[1]	1951	7 years/F	Angio done, BCV located subpulmonary, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable
12.	Sherman FE et al[38]	1963	?	Necropsy, anterior to arterial duct, left aortic arch, pulmonary atresia, VSD	Not applicable	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
13.	Sherman FE et al[38]	1963	?	Necropsy, anterior to arterial duct, left aortic arch, pulmonary atresia, VSD	Not applicable	Not applicable	Not applicable
14.	Yoshida Y et al[63]	1975	68 years/M	Necropsy, anterior to arterial duct, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable
15.	Kitamura S et al[66]	1981	69 years/M	Necropsy, anterior to arterial duct, left aortic arch, no associated anomalies	Not applicable	Not applicable	Not applicable
16.	Cloez JL et al[8]	1982	$1\frac{1}{2}$ year/M	Echo+Angio, left aortic arch, TOF	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
17.	Smallhorn JE et al[19]	1985	7 patients	Right aortic arch (6), left aortic arch (1), RVOT anomalies, "frequent: intracardiac anomalies (6), VSDs?, central pulmonary arteries absent (1)	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
18.	Townsend MD et al[13]	2008	9 years/F	Echo+Cath+Angio Diagnosis: day 8 bicuspid aortic valve, AP window repaired under DHCA, no mention of retroaortic BCV, persistent systemic arterial desaturation 9 years- hospitalized ventilated, +ve pressure, with nitric oxide. TTE- suprasystemic RVSP, CT-angio- posterior pulmonary venous drainage with a confluence draining in the LA. LSVC connected to LA appendage- LA roof junction, retroaortic BCV connected to LSVC and drained to SVC-RA junction. Saline contrast echo- bubble in LA and RA	A 5 cm long window like communication between LSVC and LAA to extend to the roof of the LA; biatrial drainage of retroaortic BCV- direct suture closed of the window	Unremarkable, SaO2 normalized.	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
19.	Towbin JA et al[12]	1987	18 months/M	Echo-TOF, high ASD, absent hepatic portion of IVC with azygous continuation, LA isomerism, RAA, retroaortic BCV (contrast echo with left hand injection), confirmed the diagnosis	Not mentioned	Not mentioned	Not mentioned
20.	Walsh R et al[29]	2017	5 years/M	Cardiology evaluation of a murmur. Echo-double aortic arch. MRI-evaluation of vascular ring and airway compression, left dominant double aortic arch with a retroaortic BCV. No airway compression	Conservative management	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
21.	Morhy Borges Leal S[23]	2002	9 months/F	Diagnosis-Echo, TOF+PS, right aortic arch, no associated anomalies, retroaortic BCV	Done	Survived	Not mentioned
			1 year 3 months/M	Echo, TOF+PS, right aortic arch, no associated anomalies, retroaortic BCV	Done	Not mentioned	Not mentioned
			2 years 1 month/F	Echo, TOF+PS, left aortic arch, no associated anomalies, , retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			7 years 5 months/F	Echo, TOF, PA atresia, right aortic arch, absent LPA, hypoplastic RPA, MAPCA's present, retroaortic BCV	Surgery done	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			1 year 1 month/F	Echo, TOF+PS, left aortic arch, associated supraval-var PS, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			7 years/F	Echo, TOF+PS, right aortic arch, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			1 month/M	Echo, TOF+PS, right aortic arch, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			8 months/F	Necropsy, TAPVC, AVSD, DORV, right isomerism, left aortic arch, Echo diagnosis	Necropsy	Not applicable	Not applicable
			20 years/M	Echo, TOF, PS, right aortic arch, no associated anomalies	Surgery done	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			36 years/F	Echo, ASD, PS, left aortic arch, no associated anomalies	Surgery done	Not mentioned	Not mentioned
			18 years/F	Echo, TOF, PS, left aortic arch	Surgery done	Not mentioned	Not mentioned
			8 months/M	Echo, TOF, PS, right aortic arch,	Surgery done	Not mentioned	Not mentioned
			15 years/F	Echo, Normal heart, left aortic arch	Not applicable	Not applicable	Not applicable
			1 month/F	Echo+ angio, TOF pulmonary atresia, left aortic arch, no associated anomalies	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
22.	Ming Z et al[35]	2009	4 months/M	Pulmonary atresia, VSD, PAD, ASD, right aortic arch, right tracheal bronchus, CT-angio, mild compression of the esophagus by retroesophageal left BCV,	Not mentioned	Not mentioned	Not mentioned
			6 months/M	Echo, VSD, CT angio-retroesophageal BCV	Not mentioned	Not mentioned	Not mentioned
			6 days/F	Pulmonary atresia, VSD, PAD, left aortic arch, CT-angio,retroesophageal left BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			6 months/F	VSD, PAD, CT angio, retroaortic BCV divided into two branches. Anterior and thinner branch was located above the aortic arch, drained into SVC; posterior thicker branch routed posterior to esophagus and joined the azygous vein before draining to RSVC	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
23.	Kim HJ et al[28]	1994	61 years/F	Sudden retrosternal pain, CXR-prominent aorta, mild widening of superior mediastinum, CT-highly enhancing venous structure lateral to aortic arch. The retroaortic BCV joined RSVC below the azygous vein; venography-confirmed the CT findings. Diagnosis of chronic liver disease	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			46 years/M	CXR-widened superior mediastinum, CT-retroaortic BCV. The vein abruptly changes its course medially at the level of APW, continued between aortic arch and lower trachea joined RSVC below azygous vein	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
24.	Elami A et al[57]	1985	6 years/F	Cyanosed TOF, CXR-classical TOF, Echo-TOF, RVOT infundibular gradient 97mmHg, absent pulmonary valve, confluent PA, RAA with mirror image branching, juxtaductal COA gradient 48 mmHg, PFO, retroaortic BCV, cath angio confirmed the diagnosis	Right thoracotomy extended resection end to end eanastomosis; 2 months later, ICR-TOF	Survived	At 12 months asymptomatic
25.	Mill MR et al[2]	1993	3 days/M	IAA, APW, VSD anomalous RSA from the DTA, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			50 months/F	TOF, right aortic arch, PFO, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
			6 months/F	TOF, right aortic arch, ASD, LSVC, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
			25 months/M	TOF, right aortic arch, Cornelia de Lange syndrome, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
			19 months/F	TOF, right aortic arch, Trisomy 21, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
			55 months/M	TOF, right aortic arch, PFO, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			24 months/M	TOF, right aortic arch, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
26.	Amerasekera SSH, McGurk SP[69]	2009	30 years/M	MRI, structurally normal heart, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned
27.	Fujimoto K et al[10]	1992	54 years/F	CXR- "bucking" of the aortic arch, suggestive of aortic aneurysm, MRI- anomalous left BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
28.	Curtil A et al[77]	1999	N=25 patients	TOF, right aortic arch (19, 70%), ultrasonographic diagnosis (19, 70%), during surgery diagnosis established (6, 22%), associated cardiomyopathy (5), malformational syndromes (2), retroesophageal subclavian artery (2), Di-Georges syndrome (1)	One stage ICR (13), systemic pulmonary shuntICR (10), not operated (2)	Not mentioned	Not mentioned
29.	Kitamura S et al[66]	1981	69 years/M	Necropsy study-retroaortic left BCV (diameter 13mm, length 7.9 cm), right SVC (diameter 15mm, length 6.7 cm)	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
30.	Chen SJ et al[78]	2005	N=30 patients (18 males)	Age: 13 days-36 years, median 1 year 7 months, left anomalous BCV (27), right anomalous BCV (1), anomalous BCV bridging between bilateral SVC (2), TOF (23), RAI (4), ASD (1), VSD (1), DORV (1), RAA (21), TOF with P atresia (10), no LSVC	Not mentioned	Not mentioned	Not mentioned
31.	Semionov A, Kosiu KJ et al[33]	2017	48 years/F	Diagnosis-carcinoma colon, no CHD, no vascular anomaly, contrast CT-normal heart and great vessels, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
32.	Kwon OH et al[47]	2014	72 years/M	Evaluation of incidental lung mass on CXR, CT chest-evaluation of pulmonary nodule, double left BCV and PLSVC, left BCV – anterior branch, normally placed left BCV; posterior branch below the aortic arch, drained in RSVC, PLSVC-connected to A via coronary sinus	Right middle lobectomy for squamous cell carcinoma	Survived	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
33.	Shaffer EW	1986	3 years/F	Evaluation of murmur, cyanosis, hypoxic spell at 5 months age, Cath-TOF, RAA, mirror image branching, left BT shunt at 5 months age, repeat cath, Echo-diagnosis confirmed, retroaortic BCV	Intracardiac repair	Not mentioned	Not mentioned
34.	Kulkarni S et al[24]	2008	0.4 years/M	Mitral atresia, hypoplastic LV, DORV, LAA, VSD, PS, normal pulmonary atresia, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			0.8 years/F	TOF, RAA, normal pulmonary arteries, retroaortic BCV	Surgery done	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
			1.5 years/M	TOF, RAA, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			16 years/F	TOF, RAA, pulmonary atresia, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			1.5 years/M	TOF, PA, post BT shunt, pulmonary atresia, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			1.5 years/M	TOF, RAA, pulmonary artery normal, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			0.4 years/F	TOF, severe PS, post BT shunt, RAA, retroaortic BCV	Surgery done	Not mentioned	Not mentioned
			0.1 years/M	TOF, sevre PS, RAA, retroaortic BCV	Surgery done	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
35.	Yigit AE et al[31]		15 years/M	Recurrent lung infection, CXR- mediastinal widening, CT angio – retroaortic BCV, divided into 2 branches at thoracic inlet. Anterior and thinner branch- above the aortic arch, anterior to CA then passed between the brachio-cephalic trunk, and LCCA- ten joined SVC, posterior thicker branch- coursed posterior to trachea + esophagus joined the azygous vein at the level of T2 and finally to RSVC, no cardio-vascular anomaly.	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
36.	Shim MS et al[46]	2010	53 years/M	Chest wall pain, CT-lung carcinoma, RUL, retroaortic BCV divided into two branches at the level of aortic arch. Anterior branch-above the aortic arch coursed anterior to the LCCA and BCA-joined SVC, posterior branch below the aortic arch, posterior to ascending aorta, both joined RSVC separately, no cardiovascular anomaly	Refused treatment	Not applicable	Not applicable

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
37.	Topcuoglu OM et al[45]	2014	62 years/M	HIV infection with cold, contrast CT-double left BCV, anterior vein normally located, posterior vein thinner, accessory branch coursing posterior to the aortic arch, anterior vein joined RSVC, accessory vein joined superior to azygous vein, diameter LBCV 12.5mm, accessory BCV 2.2 mm, RBCV 13mm	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
38.	Takada Y et al[17]	1992	6 patients M / 2 patients F	Mean age 35.7 years (range 14-58 years), CT-retroaortic BCV (6), double BCV (2), associated CHD (2), MRI same findings	Not mentioned	Not mentioned	Not mentioned
39.	Khoury NJ et al[22]	2008	53 years/M	Hypertension, hyperlipidemia, interscapular pain, MR angio+CT angio- high distal aortic arch, descending portion of the high arch- narrower in caliber for 6 cm, ascending portion- dilated, pseudo-coarctation aorta, retroaortic BCV	Not mentioned	Not mentioned	Not mentioned

S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
40.	Bartoli JM et al[55]		42 years/M	Known case of AR for 30 years, CXR LVE aortography-grade IV AR, high aortic arch, CT-high aortic arch dilated at the origin of LSA, retroaortic BCV, pseudo-coarctation aorta	AVR bio-prosthetic (CE bio 27mm)	Not mentioned	Not mentioned
41.	Subirana MI et al[43]	1986	23 years/F	Cyanotic CHD-evaluation, Echo, DORV, straddling tricuspid valve, 2 large VSDs, angio double left BCV above + below the aortic arch	Left modified BT shunt	Survived	Not mentioned

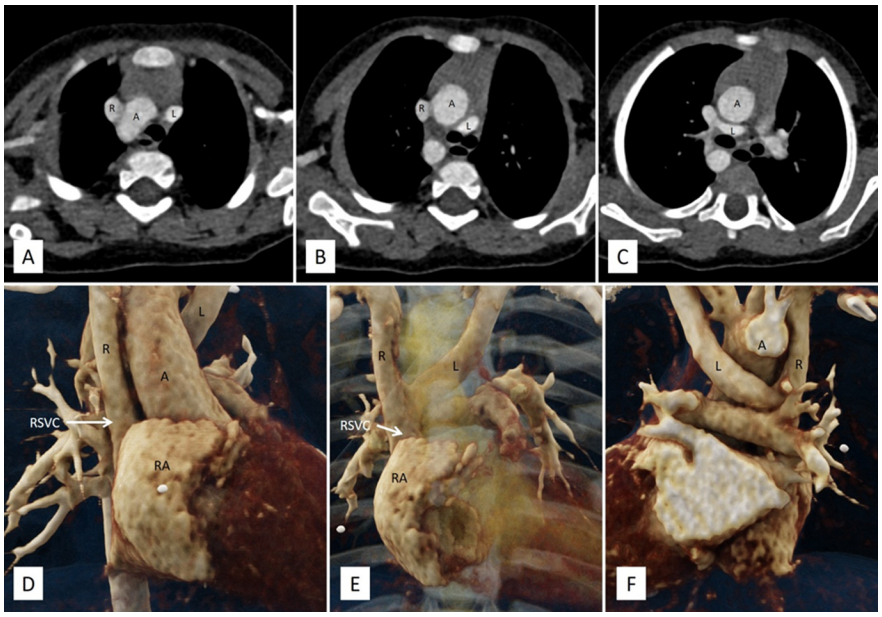
S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
42.	Koutlas TC et al[56]	1998	2 years	Diagnosis after birth-tricuspid atresia, VSD, normally related great arteries, mild deviation infundibular septum, hypoplastic RV. BAS – PA banding at 3 weeks age, Cath at 2 years-non-distorted branch PAS, mean pressure 12 mmHg, QP:QS 0.9:1, PVR 2 woods unit. Retroaortic BCV passed between LPA (anterior) and left main bronchus (posterior), 1.5 cm below the carina-compression of the left BCV between LPA and left main	CPB, circulatory arrest at 18°C, left cavopulmonary anastomosis, end to side LPA. MPA divided distal to PAB. Hemi Fontan type right cavopulmonary connection, proximal MPA to aorta anastomosis like DKS, 1 year later-fenestrated lateral tunnel Fontan	Survived	Not mentioned

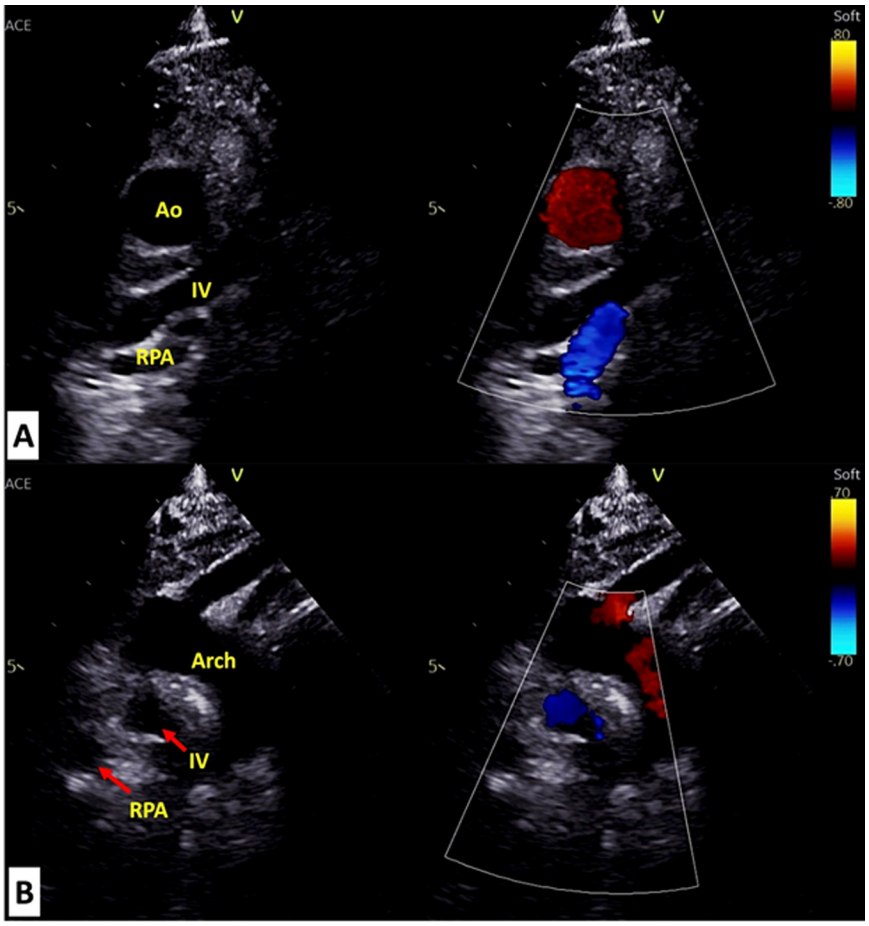
S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
43.	Agarwal R et al-74	2006	1 year 5 months	Diagnosis-tricuspid atresia, pulmonary atresia, RAA, retroaortic anomalous BCV. Day 1- LMBT, 6 months-Aorta, RPA (central shunt), 9 months-blocked central shunt, narrowed LMBT, stenting of the narrowed PAD, 1 year 5 months-narrowed LMBT, narrowed stended ductus, parallel course of the retroaortic BCV	Bi-directional Glenn, PA recon-struction. Aorta was transected for unim-peded visualization	Survived	Asymptomatic SaO2 87%

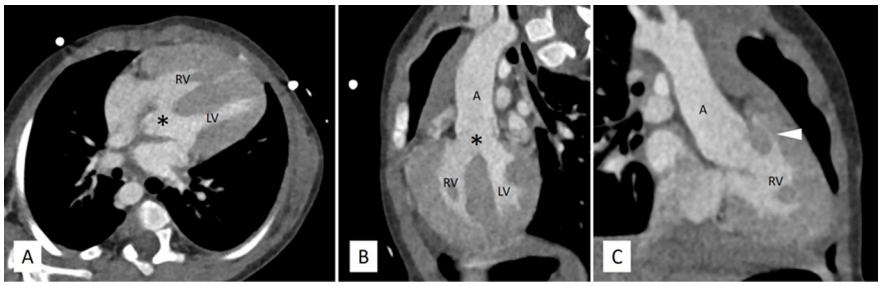
S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
44.	Konstantinov EI et al[5]	2003	New born/F	Weight 3.12 kg. Echo-COA, unrestrictive VSD, posterior malalignment outlet septum, subaortic stenosis, restrictive PFO, almost closed ductus, bicuspid aortic valve, good ventricular function, aortic annulus 0.5 cm, pulmonary annulus 1.1 cm, tricuspid annulus 1.2 cm, ascending aorta proximal to LCA 0.45 cm, juxtaductal COA, retroaortic BCV, PGE1 0.05 µg/kg/min.	CPB-bicaval cannulation, cooled to 25°C, cardioplegic arrest, PAD ligated, circulatory arrest for 25 min, ascending aorta transected – BCV transposed in front of ascending aorta, hypoplastic aorta ligated distal to LSA-ascending aorta continuity restored by a "semilunar anastomosis, descending aorta to ascending aorta anastomosis	Survived	Discharged home on 10^{th} post-operative day

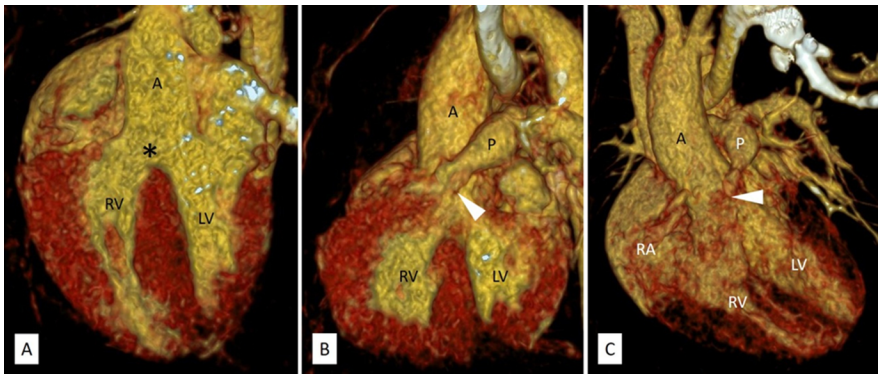
S. No.	Authors	Year	Age/Sex	Diagnosis, investigations, surgical and necropsy findings	Surgery	Results	Follow-up
45.	Nakamura Y et al[32]	2006	70 year/M	Diagnosis-post aortic left innominate vein, distal aortic arch aneurysm	Total arch replacement, antegrade cerebral perfusion at 20°C	Survived	Not mentioned
46.	Lim ZN et al[75]	2018	3 month/F	Echo, CHD, cyanosis, TOF, RAA, retroaortic BCV	CPB, intracardiac repair	Survived	At 2 months, asymptomatic
47.	Chu MW et al[56]	2007	4 weeks neonate	Echo, MR angio, respiratory distress, interrupted aortic arch, aberrant left subclavian artery, VSD, retroaortic BCV	CPB, VSD closure-Gortex patch, restoration of aortic continuity	Survived	At 3 months, asymptomatic
48.	Kawara T et al[48]	2003	3 months/M	Cath, angio, Tricuspid atresia, anomalous BCV, RVOTO, VSD, pulmonary stenosis, left aortic arch	Bilateral bidirectional Glenn without CPB	Survived	At 2 years awaiting completion Fontan

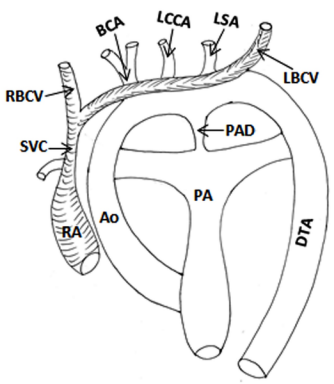
Abbreviations: AP window- Aortopulmonary window, AR- Aortic regurgitation, ASD- Atrial septal defect, AV- Atrioventricular, AVSD- Atrioventricular septal defect, BAS- Balloon atrial septostomy, BCA- Brachiocephalic artery, BCV- Brachiocephalic vein, CE bio- Carpentier-Edwards bioprosthetic valve, CHD- Congenital heart diseases, COA- Coarctation of the aorta, CPB- Cardiopulmonary bypass, CT-angio- Computed tomographic angiocardiography, CXR- Chest x-ray, DKS- Damus-Kaye-Stansel, DORV- Double outlet right ventricle, DTA- Descending thoracic aorta, Echo- Echocardiography, HIV- Human immunodeficiency virus, IAA- Interrupted aortic arch, IVC- Inferior caval vein, LA- Left atrium, LCCA- Left common carotid artery, LMBT- Left modified Blalock-Taussig shunt, LPA- Left pulmonary artery, LSA- Left subclavian artery, LSVC- Left superior caval vein, LV- Left ventricle, MAPCAs- Major aortopulmonary collateral arteries, MRI- Magnetic resonance imaging, PA- Pulmonary artery, PAD- Patent arterial duct, PFO- Patent foramen ovale, PGE1- Prostaglandin E1, PM- Postmortem, PS- Pulmonary stenosis, RA- Right atrium, RAA- Right aortic arch, RPA- Right pulmonary artery, RSVC- Right superior caval vein, RUL- Right upper lobe, RVOT- Right ventriculat outflow tract, RVOTO- Right ventricular outflow tract obstruction, RVSP- Right ventricular systolic pressure, TAPVC- Totally anomalous pulmonary venous connection, TOF- Tetralogy of Fallot, TTE- Transthoracic echocardiography, VSD- Ventricular septal defect,











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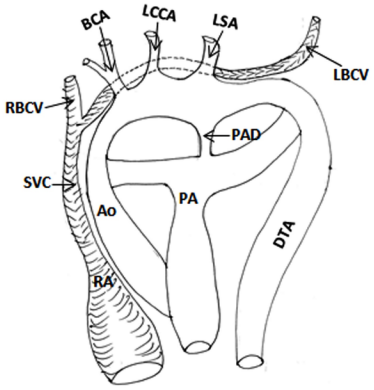

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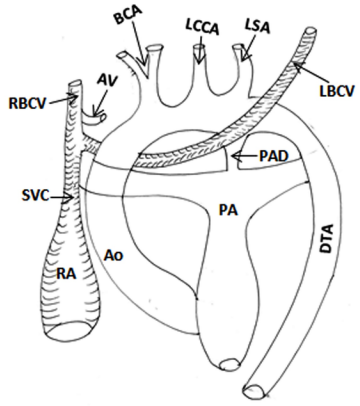

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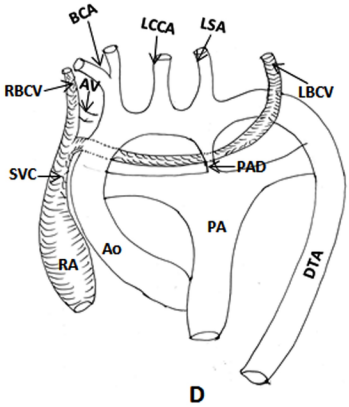

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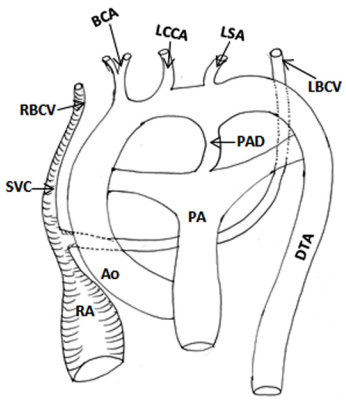

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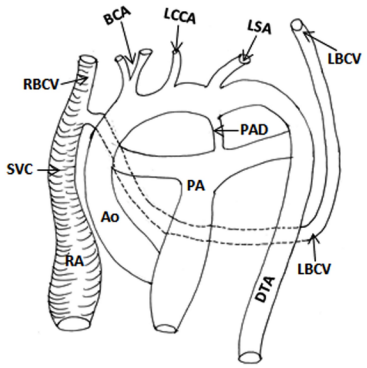

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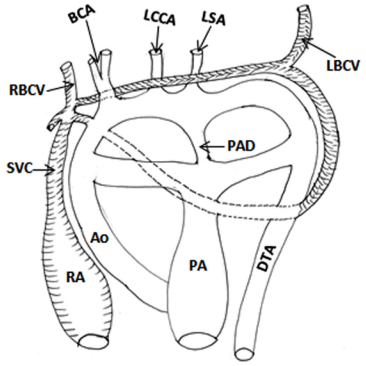

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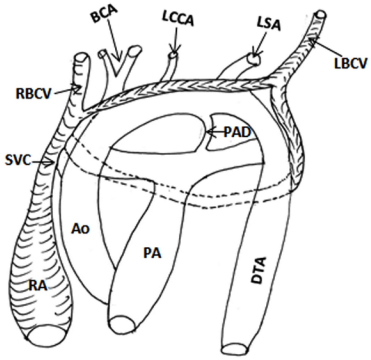

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