Surgical Management of Divided Atrial Chambers

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Abstract

The morphological variations when one, or both, of the atrial chambers is subdivided, are many and varied. We present a synthesis of 198 published investigations of this "family" of uncommon lesions. Almost three-quarters of patients with divided atrial chambers present during infancy with severe pulmonary hypertension and cardiac failure. Associated cardiac and extra-cardiac defects are present in between half and nine-tenths of cases. Acquired division of the left atrium has been reported after the Fontan operation, orthotopic cardiac transplantation, and complicated aortic valvar infective endocarditis. Surgery under cardiopulmonary bypass remains the definitive treatment. Balloon dilation may be considered in anatomically compatible variants in the setting of cardiac failure and pregnancy as a bridge to definitive treatment. Overall, mortality has been cited between nil to 29%. Presentation during infancy, associated congenital anomalies, pulmonary hypertension, and surgery in the previous era, have been the reported causes of death. The operative survivors have long-term favourable outcomes, with near normal cardiac dimensions and low risk of recurrence. While asymptomatic patients with division of the right atrium do not need treatment, surgical resection of the dividing partition under cardiopulmonary bypass is recommended in symptomatic patients with complex anatomy, the spinnaker malformation, or associated cardiac anomalies. Balloon dilation may be considered in uncomplicated patients with less obstructive lesions. Hybrid intervention and endoscopic robotic correction also have been performed. We submit that an increased appreciation of the anatomic background to division of the atrial chambers will contribute to improved surgical management.

Introduction

Sub-division of one or other of the atrial chambers is an exceedingly rare congenital cardiac malformation. Understanding to date has been confused by the on-going suggestion that such subdivision might produce three atrial chambers. As we will show, this is never the case. The lesions are properly analysed on the basis of subdivision of the existing atriums (Figures 1A-1C).

Division of the left atrium was first described by Church in 1868,[1] albeit that the anatomical arrangement had previously been illustrated by Andral in 1829.[2] It was Borst, in 1905, who introduced the inappropriate and confusing term of "cor triatriatum sinister".[3] Most cases reported over the subsequent decades were described as individual entities, but in 1959 Niwayama, in reviewing the previous literature, provided necropsy data for 4 cases.[4] Then, in 1969, Van Praagh and Corsini speculated on the morphogenesis, having assessed 13 post-mortem specimens.[5] The first diagnosis based on angiography had been made by Miller in 1964,[6] while diagnosis on the basis of echocardiographic interrogation was reported by Ostman-Smith and her colleagues in 1984.[7]

Already in 1956, surgical correction had been achieved by Vineberg, who used his finger inserted through the left atrial appendage to fracture the dividing partition.[8] Lewis and colleagues, in the same year, described their experience incising the fibromuscular shelf using hypothermia and inflow occlusion. [9] Up until 2019, we

have been able to identify around 435 publications on division of the left atrium.[10-15] Acquired division can occur, but is uncommon. We have discovered six case reports following orthotopic cardiac transplantation, an extracardiac Fontan operation, and complicated aortic valvar infective endocarditis.[16-21]

Division of the morphologically right atrium by persistence of the valves of the embryonic systemic venous sinus was initially reported on the basis of autopsy findings. [22] A clinically diagnosed case was described by Verel, Pilcher and Hynes in 1970, while successful surgical correction was reported by Hansing, Young, and Row in 1972.[E101-E104] In 1983, Mazzucco and associates reported the first successful correction during infancy.[E105] Then, in 1989, Goldfarb and associates reported sub-division of both atrial chambers.[E106]

Division of the morphologically left atrium has been estimated to be around 0.1% to 0.4%, while persistence of the valves of the systemic venous sinus accounts for 0.025%-0.1% of congenital heart diseases.[22-24,E107] Associated congenital and acquired cardiac lesions have been reported in one-third to three-quarters of those with divided left atriums, and in one-half of those in whom the morphologically right atrium is divided.[E101-E168]Limited information is available, however, on the short- and long-term outcomes of treatment for these rare conditions.

In this narrative review, we discuss the anatomical details, diagnostic challenges, associated cardiac anomalies, therapeutic techniques, and their outcomes, including reinterventions.

Methods

We searched the literature for the described instances of triatrial hearts, cor triatriatum sinister, congenital division of left atrium, cor polyatriatum, divided left atrium, subdivided left atrium, triatrial hearts, and cor triatriatum dexter. We then collated and analysed the anatomical descriptions, presence of associated cardiac and non-cardiac anomalies, including description of so-called "heterotaxy", paying special attention to surgical and non-surgical treatments used 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.

On this basis, it proved possible to make an individualized review of 198 investigations.[1-30,E1-E168] These findings were then incorporated, as far as possible, with the results of a systematic review of cases presenting during adulthood, emanating from Copenhagen, which itself had collated the findings from 171 published cases.[E57] We then synthesized the overall clinical and autopsy data to identify any issues of concern, any clues to either pre- and postnatal diagnosis, and the therapeutic options for repair of the primary as well as concomitant anomalies, hoping to improve future surgical management. Due to limited sample sizes, the heterogeneity of clinical state at the time of surgical intervention, and the difficulties in selection of appropriate cardiac quantifiable end points, it did not prove possible to perform a meta-analysis. We have sought to collate the overall findings in Tables E1 and E2.

Results

Demographics

The age of those with division of the morphologically left atrium ranged from 1 day to 93 years. In surgical series, we found a median age of 21 months, with a range from 1 to 228 months. In case reports and case series, in contrast, the median age was16 years, with a range from 1 year to 93 years.[1-30,E1-E100] In the analysis from Copenhagen, 171 individuals were identified having been diagnosed in adulthood.[E57] Their median age at diagnosis was 43 years, with an interquartile range from 30 to 60 years. Among the overall group of patients, two-thirds were male, with no identifiable regional or ethnic predominance. There were no reports of so-called "heterotaxy", including bronchial isomerism, nor any incidences of familial occurrence (Supplemental Tables E1 and E2). Almost three-quarters of described patients presented during the neonatal period, infancy, or early childhood, with shock, cardiac failure, and varying grades of pulmonary hypertension.[4,E1-E12]

The diagnosis had been an incidental finding in almost one-fifth of the 171 adults reviewed in the analysis from

Copenhagen.[E57] Of these adults, two-fifths with obstructive physiology had higher occurrences of congestive cardiac failure, pulmonary hypertension, hemorrhagic episodes, variceal bleeding, infections, and sudden death. An acute presentation with onset of symptoms within one month of the diagnosis had been observed in one-fifth. Within these adults, thromboembolic and ischemic events of various forms had occurred in one-sixth.[4,E1-E12,E69-E80] Diagnosis in the overall group had been made during pregnancy in 10 patients, and in 1 after successful labour.[E81-E91]

Asymptomatic patients were diagnosed incidentally, with the great majority having cardiac murmurs.[E4,E13,E14] The precipitating factors for appearance of symptoms could have been fibrosis and calcification of orifice within the dividing partition, development of mitral regurgitation, or development of atrial fibrillation.[28,E1-E4,E12-E15]

Those individuals with division of the morphologically right atrium were diagnosed between the ages of 1 day and 86 years, with a median of 23 years.[E101-E168] Among these individuals, seven-tenths were female. Clinical presentation depended on the severity of obstruction to systemic venous flow, and the patency of the oval foramen. Individuals with non-obstructive lesions and an intact interatrial septum may remain asymptomatic, presenting as the so-called Budd-Chiari syndrome, or with supraventricular arrhythmias.[E16-E18,E58,E65,E117,E167] When there is an interatrial communication, patients may have varying degree of intermittent cyanosis, ranging from severe hypoxia at birth to cyanosis in adulthood.[E101-E103,E105,E111,E118-E120,E122-E124]

Diagnosis

When making the diagnosis, it is desirable to have detailed anatomical information on the type and location of the dividing interatrial shelf, since the optimal surgical strategy depends on the morphology of subdivision, along with the nature of concomitant cardiac anomalies if present. In this regard, there has been a marked shift in the diagnostic modalities used over the years. In the early years, angiography, surgical observation, or autopsy had provided the arbitrating diagnostic criterions. Despite improvements in angiographic techniques, nonetheless, subdivision of the left atrium proved frequently to have been overlooked or misdiagnosed. This was because the partition could produce features of mitral stenosis, a supramitral ring, primary pulmonary hypertension, or progressive pulmonary pathologies which might cause pulmonary hypertension.[7,11,28,E4,E12,E13,E15-E18]

Since the late 1980s, either cross-sectional or three-dimensional echocardiography, including transesophageal imaging, have emerged as superior diagnostic modalities. They provide better images of the left atrium, its appendage, the morphology of the dividing shelf, including the size, location, and number of fenestrations, its spatial relationship with septal defects; and the resulting degree of obstruction. Postoperative echocardiographic interrogation then permits assessment of the adequacy of surgical treatment (Figures 2-5).[7,E12,E15,E17-E19,E21-E28]

In typical cases, such interrogation reveals the presence of a thin, linear partition dividing the left atrium into pulmonary venous and vestibular compartments. The shelf may move throughout the cardiac cycle. On the basis of its relationship to the mouth of the appendage, it can readily be distinguished from the supramitral ring[7]. The addition of Doppler technology permits estimation of the gradient between the atrial compartments.[7,E12,E15,E17-E19,E21-E28] The addition of cardiac computed tomography, or magnetic resonance imaging, serves to enhance recognition of the anatomical details (Figures 6A-6C, 7A-7C).[E2,E5-E9,E29-E32] Cardiac catheterisation with selective pulmonary angiography may still be indicated in doubtful cases, and can be used to assess pulmonary vascular hemodynamic. Elevated pulmonary arterial and pulmonary capillary wedge pressures in presence of normal left ventricular end-diastolic pressures are indicative of left ventricular inflow obstruction.[E2]

The diagnosis of division of the morphologically right atrium requires a high index of suspicion, together with thorough echocardiographic imaging. Our analysis revealed an isolated case report of prenatal diagnosis.[E121] The entity certainly belongs to the differential diagnosis of neonatal cyanosis.[E122-E125]On M-mode echocardiography, the lesion can present as a cloud of echoes penetrating the right ventricle during diastole. Due to

improved diagnostic accuracy with saline contrast echocardiography, there is now an increase in its reported frequency.[E126-E132]Magnetic resonance imaging is now recognised as a modality with which to delineate the arrangement of the persisting valves of the systemic venous sinus. It provides the gold standard for assessment of ventricular volumes.[E130,E131]

Clues to diagnosis in those with divided left atrium can be provided by chest radiography, which typically shows cardiomegaly with right ventricular enlargement, occasional left atrial enlargement, and pulmonary congestion. Kerley B and C lines are frequently found. Electrocardiographic evidence of right ventricular hypertrophy, right axis deviation and an S_1, Q_3T_3 pattern is common. The overall findings in the presence of divided right atrium can be characterized in terms of central cyanosis, clubbing, bilateral lower limb edema, clear lung fields, a widely split second heart sound hepatic enzyme abnormalities, and right bundle branch block.

Surgical Anatomy

The essence of division of the morphologically left atrium is the presence of a fibromuscular shelf that courses diagonally so as to divide the chamber into pulmonary venous and vestibular compartments (Figures 1A-1C). In a small minority of cases, the chamber can be divided into a vestibular compartment and a blind second part, with the pulmonary veins connects in totally anomalous fashion. In another very small minority, the vestibular chamber can be blind in the setting of mitral atresia. It is the presence of the mouth of the left atrial appendage in the vestibular chamber that distinguishes the lesion from the supravalvar mitral shelf.[**13,E51**] The dividing shelf itself is usually incomplete or fenestrated, but can on occasion be imperforate. Its size, shape, thickness, and location vary markedly from patient to patient. Many schemes have been proposed to classify the malformation.[10,23,27,E18,E33,E34] The simplest approach is to recognise that the partition does no more than divide the left atrium into pulmonary venous and vestibular compartments.[E51,E52,E108] The important additional feature is the site of the oval fossa, as was emphasized in the reference by Bharucha and colleagues.[E71] The very cases not fitting these patterns typically reflect the presence of associated anomalous pulmonary venous connections, either total or partial.[23,27,E33,E34,E71] It is also necessary then to document whether an interatrial communication communicates with the pulmonary venous, or distal, compartment, or with the vestibular chamber. Note should obviously also be taken of the presence and size of the communication between the atrial compartments, but once diagnosed, this feature will not influence the surgical repair (Figures 1A-1C, 6A-6C, 7A-7C). Details of the previous categorisations are now of only historical interest.[23,27,E33,E34] The simplified approach of recognising pulmonary venous and vestibular chambers must, of course, exclude those individuals with supravalvar or intravalvar stenosing mitral rings.[23,27,E33,E34] This simple classification can also incorporate equally rare cases reported in which dual or triple partitions have produced still further subdivision of the left atrium. It does not help understanding to describe such rare lesions as "cor polyatriatum", just as it no longer helps to describe the divided left atrium as representing "cor triatriatum sinister".[E94-E96]

Associated cardiac lesions

One-third to three-quarters of the reported individuals with divided left atrium had associated congenital or acquired anomalies.[11,E35-E44] Patency of the oval foramen, or a defect in the oval fossa, were the most common concomitant defects. Other cardiac anomalies included partially anomalous pulmonary venous connection, persistent left superior caval vein, persistent patency of arterial duct, pulmonary venous stenosis, superior sinus venosus defect, common atrium, atrioventricular septal defect, ventricular septal defect, tetralogy of Fallot, pulmonary valvar or infundibular stenosis, pulmonary arterial stenosis, right-sided heart, mirror-imagery, transposition, hypoplastic pulmonary trunk, double orifice mitral valve, unroofed coronary sinus, hypoplastic left heart syndrome, tricuspid regurgitation, myxomatous mitral valve disease, degenerative aortic stenosis, Wolff Parkinson White syndrome and restricted mitral valve disease.[11,E35-E44]

Associated cardiac anomalies in adults include mitral regurgitation, atrial septal defect, persistent left superior caval vein, unroofed coronary sinus, aortic regurgitation with dissecting aneurysm, tricuspid regurgitation, supravalvar mitral ring / stenosis, mitral valve stenosis / hypoplasia, parachute mitral valve, par-

tially anomalous pulmonary venous connection to brachiocephalic vein, right atrium, superior cavoatrial junction, or right superior caval vein; bicuspid aortic valve, coarctation of aorta, aberrant right subclavian artery, hypoplastic left heart, tricuspid stenosis / atresia, subaortic stenosis and valvar pulmonary stenosis.[E4,E5,E12-E14,E37]

According to Hansing, Young, and Row, diagnosis of the divided right atrium should be restricted to individuals in whom the valves of the embryonic systemic venous sinus have persisted to the extent that they interfere with normal systemic flow in the right atrium.[E52,E111] Persistence of the venous valves can retard the growth of the remainder of the right atrium, often being seen in the setting of tricuspid atresia.[E101,E102,E111,E113,E133,E149] In some cases, the persistent valve expands to form a spinnaker, or windsock-like, structure consisting of excess fibrous tissue with a thin, distensible centre and cord-like attachments along the parietal wall of the right atrium. The structure can prolapse into the right ventricle, and out through the right ventricular outflow tract and pulmonary trunk.[E111,E113-E115,E133,E134] Associated right-sided congenital cardiac anomalies are present in almost half of the reported cases. They include hypoplasia or atresia of tricuspid and/or pulmonary arterial orifice, Ebstein's malformation, tetralogy of Fallot, right ventricular hypoplasia, and transposition.[E101-E105,E111-E113,E119,E120,E126,E130,E134-E141]There are isolated reports of coarctation of aorta, hypoplastic left heart syndrome, and right atrial thrombus.[E142,E143]

Surgical Approach and Management

For those with divided left atriums, medical management alone is ineffective. It should be used only in incidentally diagnosed asymptomatic patients, and to control supraventricular arrhythmias. It should be recommended only when the opening within the dividing partition is non-restrictive, and regular echocardiographic follow-up is possible.[E45] Most reported patients were symptomatic early during infancy, and underwent rapid surgical correction.[24,E1-E5,E12-E16,E37] Among 171 reported adults, however, almost half did not require any intervention.[24,E1-E5,E12-E16,E37,E57] It follows that those diagnosed with obstructive symptoms at an early age should undergo surgical correction, with the urgency of operation primarily determined by the severity of the presenting symptoms.

Surgical management has evolved with time. Moderately hypothermic cardiopulmonary bypass at 32°C with cold cardioplegia is now the most popular technique. Deep hypothermic circulatory arrest has been employed when separate cannulation of the caval veins appeared to be impractical because of venous anomalies and the small size of the operating field.[10-15,E105] The procedure requires no more than complete excision of the dividing shelf so as to provide unrestricted antegrade pulmonary venous blood flow. It is, of course, also necessary to take care of concomitant cardiac anomalies.[10-15,E35,E36,E43,E44,E150]

The dividing shelf can be approached from either a left or right atriotomy, depending on the presence of an atrial septal defect.[10-15,E35,E36,E43,E44,E150] Any complicating anatomical features should have been identified before proceeding to correction. On this basis, it is desirable to approach through the largest atrial chamber. Having excised the shelf, it may be necessary to reconstruct the atrial septum with a pericardial baffle or patch lest there be a residual defect. Transcatheter balloon dilation has been successfully performed in pregnant individuals in cardiac failure, where conventional cardiopulmonary bypass is associated with higher perioperative mortality and morbidity.[E38-E42] There are now isolated reports appearing of hybrid intervention and endoscopic robotic correction.[E57,E92]

Asymptomatic patients with divided right atrium, and limited or no obstruction, usually do not require any intervention.[E45] If the patient is undergoing cardiac surgery for other reasons, the persisting venous valves can be removed as part of the original procedure. Management of symptomatic individuals depends on the severity of obstruction. Surgical resection of the venous valves under cardiopulmonary bypass, particularly if forming a spinnaker-like malformation, is the treatment of choice for symptomatic patients with significant right ventricular inflow obstruction, cyanosis, complex anatomy, and associated congenital heart diseases.[E105] There are isolated reports of successful transcatheter balloon dilation, but this can only be employed in patients with less obstructive lesions with uncomplicated anatomy.[E143-E145] Percutaneous catheter-based disruption of the venous valves has been reported when identified as forming a Chiari network, although

the redundant network makes catheterisation difficult because of difficult navigation or entrapment of the sheaths and/or the closure of an associated interatrial communication.[E143-E148, E164,E165]

Short- and Long-term Follow-up

Death after surgical repair of divided left atrium is now uncommon, and usually related to accompanying complex congenital cardiac malformations, year of operation, and the severity of pulmonary hypertension. Mortality rates in older series, including both children and adults undergoing surgery between 1959 to 1992, ranged from 8% to 29%. These had decreased to zero to 4% in more recent reports.[10-14,E18,E42-E44,E101,E102]

Saxena and associates from Mayo Clinic reported 25 surgical patients with 83% long-term survival and no instances of recurrent pulmonary vein stenosis or recurrent divided left atrium at a median follow-up of 6.6 years.[E44] During a median follow-up of 5.4 years of a cohort of 65 patients from Boston Children's Hospital, none required re-intervention for recurrent left atrial obstruction, while six of eight patients with pulmonary vein stenosis underwent re-intervention.[E43]

Alphonso and associates from Melbourne Children Hospital reported 28 surgical patients with no requirement of reoperation for recurrent pulmonary vein stenosis or recurrent left atrial obstruction at a median follow-up of 98 months.[E36] Humpl and associates from Toronto reported 82 patients diagnosed between 1954 and 2005, of which 19 (23%) died at a median of 2 months after presentation. Operative mortality was 9%. Kaplan-Meier survival was 86% at 5 years. There was no change in mitral valve Z-score over time.[E35]

In the systematic review of 171 adults with divided left atrium, 71 (41.5%) patients required interventional treatment [surgical resection of the membrane (65), percutaneous balloon dilation (3), hybrid intervention (2), and endoscopic robotic correction (1)]; seventy-eight (45.6%) patients not requiring any intervention, and 12 (7%) refusing treatment.[E57-E71]

Surgical resection of partition producing division of the right atrium is curative.[10-14,E1,E2,E18,E42-E44] Successful percutaneous balloon valvuloplasty of subpulmonic membrane has been reported as a bridge to definitive surgical treatment.[E145] Spontaneous involution of Chiari network with moderate right ventricle inflow obstruction has also been reported in early infancy.[E148]

Discussion

The description of division of the atrial chambers, across the recent decades, has been confused by suggestion that the lesion produces three such chambers. This is never the case. There has never been a case reported, of which we are aware, in which it is possible to recognise three morphologically distinct atriums. Instead, the lesions are simplified by the recognition that either the morphologically right or morphologically left atrium can be congenitally divided.[E51,E108,E109] Such division, nonetheless, remains rare. As yet, there has been no convincing account provided to explain division of the morphologically left atrium, but division of the morphologically right atrium is readily explained on the basis of persistence of the valves of the developing systemic venous sinus.[22,E52] It is incomplete involution of these valves that accounts for the spectrum of anatomical presentations. Persistence of the right valve in the simplest form appears as a prominent or giant Eustachian valve.[E111] Chiari network represents a more incomplete involution, presenting in up to one-twentieth of the population.[E110,E114,E119] Divided right atrium is no more than the more severe form, with no or minimal involution of tissues of the venous valves.[E111,E112]In the most severe cases, the tissue forms a windsock spinnaker-like membrane across the tricuspid valve, which causes severe right ventricular outflow tract obstruction.[E111,E112,E114,E115,E133] In severely obstructive cases, bipartite right ventricle, pulmonary atresia, and tricuspid atresia have been reported.[E111-E115]

The natural history of divided left atrium depends mainly on the size of the hole in the dividing shelf, the presence and location of interatrial communications, hemodynamics, and associated congenital cardiac anomalies.[12,E35,E37,E45] Although patients may be asymptomatic, two-thirds of those presenting in infancy are shocked, have pulmonary edema, respiratory failure, and pulmonary hypertension. About three-quarters of those born with the classical lesion were previously reported to die in

infancy.[E37,E45]Asymptomatic patients without obstruction, in contrast, have a benign prognosis and present in adulthood.[13,24,E1-E5,E12-E14,E16,E17,E45] We were unable to find any account of progressive obstruction.

The presence of associated defects can sometimes mask the presence of divided atrial chambers, particularly when the arrangement itself is atypical.[E50] For example, we discovered 10 reports of division of the morphologically left atrium in the setting of virtual absence of the atrial septum. This is often termed "common atrium", which is not incorrect. The finding of the shelf does not mask the fact that there remain two atrial chambers. When the morphologically left atrium is divided in this setting, however, pulmonary venous obstruction can occur subsequent to surgical septation should the dividing shelf be overlooked.[E96-E100] In this regard, any lesion that causes intracardiac stasis over several years can pose a cardioembolic risk. It is not surprising, therefore, that an important presentation of divided left atrium in adults is cardioembolic stroke. At least 27 such cases have been reported.[E57,E72-E80] It is always necessary, therefore, to exclude associated anomalies once the diagnosis of divided left atrium has been made. As we have emphasised, the frequent lesions are an atrial septal defect, and persistence of the left superior caval vein.[7,11,14,15,24,30,E2,E35-E37,E42-E45,E49] Totally or partially anomalous pulmonary venous connections are particularly important. In the past, totally anomalous pulmonary venous connection to the coronary sinus was incorrectly interpreted as producing a triatrial heart in its own right. Such totally anomalous connection can be found when the remainder of the left atrium is divided, but the compartment usually receiving the pulmonary veins will then be blind ending. Association with pulmonary venous stenosis can also occur, and has been reported in up to one-tenth of cases at the time of repair, and in one-twentieth following corrective surgery.[E43,E44,E46]Of those with recurrent pulmonary venous stenosis, however, none had recurrence of the atrial division, and all were alive at the time of detection. This suggests that the pulmonary venous disease in this setting is not as malignant as the progressive stenosis observed following repair of totally anomalous pulmonary venous connection.[10-14,E1,E2,E18,E42-E44]

Associated abnormalities of the mitral valve occur in up to one-quarter of patient, including clefting, hypoplasia, stenosis, and the parachute lesion.[E4,E5,E12-E16,E37] Mitral valvar atresia is also reported. In this instance, the vestibular compartment of the divided atrium will be blind-ending. In three case series, supravalvar stenosis is reported along with division of the left atrium.[4,E43,E44] This is another instance where the heart might be described, inappropriately, as having four atrial chambers.[E94-E96]

As we have already emphasised, persistence of the left superior caval vein is one of the commonest associated anomalies.[28]Indeed, some have sought to invoke the presence of the persistent caval vein as a trigger to production of the dividing shelf. If this was the case, however, then all individuals with the divided left atrium would be expected also to have persistence of the left caval vein. When it is found, almost always the vein drains to the right atrium through the coronary sinus. In the minority, there is unroofing of the coronary sinus, a feature of obvious surgical significance.[10-15,E35,E36,E43-E48]

The diagnosis should always be suspected in patients presenting with pulmonary hypertension or pulmonary venous obstruction. Presence of the divided left atrium can also mimic other diseases, such as mitral stenosis, supramitral ring, primary pulmonary hypertension, and those progressive pulmonary conditions causing pulmonary hypertension. This has resulted in misdiagnosis in multiple instances.[7,10,11,13,28,E16,E21,E36,E46]

Unlike division of the left atrium, which carries high mortality if not repaired, division of the morphologically right atrium has diverse clinical presentations. They depend on the degree of partitioning of the right atrium, the patency of atrial septum, and the extent of any obstruction to flow. In the absence of an interatrial communication, and with no obstruction to flow, patients may remain asymptomatic, with the lesion discovered as a chance finding.[E101-E165] When the atrial septum is patent, individuals may present with varying degrees of intermittent cyanosis, syncope, paradoxical systemic and coronary embolism ranging from severe hypoxia after birth, to cyanosis in childhood.[E101-E103,E105,E118-E120,E112-E125] The differential diagnosis includes flail tricuspid valvar leaflet, small right heart thrombus, or a pedunculated right atrial myxoma. As we emphasised, echocardiography is now the usual diagnostic modality.[E126-E129] Magnetic resonance imaging, nonetheless, has been shown ideal for delineating the anatomy in complex cases, and is the gold standard for assessment of ventricular volumes. The latter feature can be crucial prior to anticipated surgical intervention, particularly if it is necessary to assess the feasibility of one and one-half ventricular as opposed to biventricular

repair.[E130,E131] Surgery remains the gold standard for management of symptomatic individuals. The reported operative mortalities have ranged from zero to 29%. The higher mortalities are obviously found in older series, but can still be found in those with associated complex congenital heart diseases, those presenting in infancy with cardiac failure, or when there is severe pulmonary hypertension.[10,11,13-15,E35,E36,E42,E44] An association with complex additional malformations adversely affects not only early but also late survival.[11,13-15,24,E44] We were unable to find any reports of recurrence following surgical resection.[E147,E148]

Acquired division of an atrial chamber is particularly uncommon. It has been reported, nonetheless, subsequent to the Fontan operation, orthotopic cardiac transplantation, or in the setting of complicated aortic valvar infective endocarditis in adults. The dividing partitions in these instances were produced by hypertrophied atrial tissue, suture lines, torsion of the atrial walls, or infolding of the redundant donor atrial tissue.[16-21]

Conclusion

We can conclude, on the basis of our review, that division of one or other of the atrial chambers is an uncommon congenital cardiac malformation. Understanding to date has been confounded by the suggestion that such lesions produce three atrial chambers. This is not the case. Instead, one or other of the atrial chambers is divided into two or more compartments. Multimodality imaging is now able to characterize, delineate, and differentiate such atrial subdivision from other cardiac anomalies. Despite the diversity in presentations and complexity of associated lesions, resection of the dividing shelf allows the operative survivors to regain near normal cardiac dimensions, producing a long-term favourable outcome with a low risk of recurrence.

Author's contribution

Author's name	Concept/ design	Data analysis/ interpretation	Drafting article	Critical revisi
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Figure Legends

Figures 1A-1C: The anatomical images show the essence of subdivision of the atrial chambers. Panel A shows the typical arrangement in which a diagonal shelf divided the morphologically left atrium into pulmonary and vestibular chambers. The oval fossa in this example is in communication with the pulmonary venous compartment. Panel B shows the variant in which the oval fossa, in this instance deficient, is opening into the vestibular compartment. Rarer variants can be associated with totally anomalous pulmonary venous connection, or mitral atresia, in which instance one of the compartments will be "blind-ending". Panel C shows the feature of division of the morphologically right atrium, which is persistence of the valves of the embryonic systemic venous sinus. In this example there is associated tricuspid atresia. Analysis on the basis of division of the atrial chambers resolves the complications that ensue when classifications have been attempted, incorrectly, on the presumption that there are three atrial chambers.

Figure 2: The preoperative two-dimensional transthoracic echocardiogram of 4-chamber apical view shows the divided left atrium into pulmonary (PC) and vestibular chambers (VC) by a dividing shelf (thick arrow) under mitral valve (dashed arrow). There is a prominent moderator band (thin arrow) in the enlarged right ventricle (RV).

Figure 3: The postoperative transthoracic echocardiogram, with colour Doppler interrogation shows laminar flow of blood across the left atrial outflow (red arrow). LA, left atrium; LV, left ventricle; RA, right atrium; RV, right ventricle

Figure 4: The postoperative transthoracic echocardiogram shows the patch used in this patient to route a ventricular septal defect to the aorta (red arrow). A small residual portion of a pre-existing shelf can still be visualized in the left atrium (red dashed arrow). It was not causing any obstruction to the left atrial outflow

Figure 5: Continuous wave Doppler interrogation across the tricuspid valve revealed moderate tricuspid regurgitation with a peak velocity of 3.07 m/s

Figure 6A-6C: Volume rendered images (A to C) reveal a thick shelf-like partition (yellow arrows in A and B; shaded yellow structure in C) dividing the left atrium into two chambers, a postero-superior pulmonary venous chamber which receives the pulmonary veins (PV), and an antero-inferior vestibular chamber which communicates with the left atrial appendage (LAA) and the mitral valve (MV). The two chambers communicate across a fenestration in the dividing shelf of good size (curved arrow in A and B; asterisk in C).

Figure 7A-7C: Four chamber reconstruction (A) and volume rendered image (B) of CT angiography demonstrating a thick shelf-like partition (black arrows in A and B) that divides the left atrium into postero-superior pulmonary venous chamber receiving the pulmonary veins (PV) and an antero-inferior vestibular chamber which communicates with the left atrial appendage (LAA) and the mitral valve (MV). Volume rendered image (C) showing the two chambers communicating across a good-sized fenestration (indicated by arrowheads) in the dividing shelf.

Table E1: Summary of the published investigations documenting the diagnosis of divided left atrium with or without associated congenital cardiac anomalies and its management

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
1.	Arciniegas E et al, 1981[10]	1971-1980	6	Mean 22 months, infants 3 (50%), oldest 93 years	Asymptomatic 3 (50%), Echo (0), Cath (6, 100%), Correct pre op. diagnosis (11, 56%), associated anomalies (atypical including LSVC (3, 50%), oval fossa-PC, no ASD (1); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (5)	No. 6, Surgical Approach; RA (6), DHCA (2)	HD (1, 16%), mean follow-up 2 years, LD (0), reoperation (0)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
2.	Richardson JV et al, 1981[12]		21	Mean 26 months, infants 12 (57%), oldest 56 years	Asymptomatic 2 (10), Echo (0), Cath (18, 86%), Correct Pre op. diagnosis (9, 43%), Associated anomalies (atypical-5, 24%), oval fossa-PC, no ASD (1); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (20)	Surgery (14), Surgical Approach: RA (9), LA (5) DHCA (5)	HD (6, 29%), atypical (1), mean follow-up 3.5 years, LD (1) re-operation (1) for pulmonary vein stenosis

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
3.	Oglietti J et al, 1983[11]	1959-1980	25	Infants (8, 32%), oldest 38 years, Females, 12	Asymptomatic (0), Cath (25, 100%), Echo (0), Correct pre op. diagnosis (14, 56%), associated lesions-atypical (20) oval fossa-PC, no ASD (5); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (15)	Surgery (25), Approach-RA (12), LA (10), biatrial (3), DHCA (0)	HD (4, 16%, atypical=4), mean follow-up-unknown, reoperation (1)- for residual cor triatriatum

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
4.	Rodefeld MD et al, 1990[13]	1979-1989	12	Mean age 37 months, infants (5, 45%), oldest 15 years Males, 6	Asymptomatic (0), Cath (11, 92%), Echo (12), Correct pre op. diagnosis (12, 100%), associated anomalies atypical (6, 50%), oval fossa-PC, no ASD (7); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (3)	Surgery (11), Approach-RA (6), LA (4), biatrial (2), DHCA (0)	HD (2, 16%), atypical (2), mean follow-up 1.8 years, LD (0), reoperation (0)
5.	Gheissari et al, 1992[E47]	1960-1988	12	Mean age 11 months, oldest 7.5 years, Sex; NA	Asymptomatic (0), Cath (6, 50%), Echo (3, 25%), Correct pre op. diagnosis (9, 75%), associated anomalies (atypical; 9, 75%)	Surgery (7), Approach RA (6), LA (1), DHCA (0)	HD (1, 8%), mean follow-up, reoperation, LD-unknown

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
6.	Salomne G et al, 1991[14]	1973-1989	15	Mean age 75 months, infants 8 (53%), oldest 48.1 year	Asymptomatic (2, 13%), Cath (11, 73%), Echo (4, 31%), Correct pre op. diagnosis (12, 80%), associated anomalies (4, 27%), oval fossa-PC, no ASD (5); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (10)	Surgery (15) approach-RA (13), LA (2), DHCA (6)	HD (3, 20%), mean follow-up (4.8 years), LD, reoperation-nil

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
7.	Van Son JA et al, 1993[15]	1960-1992	13	Mean age 9.4 months, infant (1, 7%), oldest patient 57 years	Asymptomatic (4), Cath (10), Echo (3), Correct pre op. diagnosis (11, 85%), associated anomalies (atypical) (6, 46%), oval fossa-PC, no ASD (4); oval fossa-PC, ASD between PC and RA, ASD between VC and RA (3)	Surgery (13), RA (6), LA (7) DHCA (1)	HD (1, 7%), mean follow-up 15.7 years, LD (1), reoperation (1)-pulmonary artery
8.	Alphonso N et al, 2005[E36]	1981-2003	28	Median age 6 months (0.6-240 months), >5 years n=4, <1 year (15), Neonates (7), infants (8), females, 15	Echo (27-96%), Cath (9-32%)-all with atypical CT, correct pre op. diagnosis (27, 96%)	Surgery (27), RA (26), LA (1)	HD (1), median follow-up 98 months (0.2-284 months), LD (1-10 years), post repair survival 96% and 88% at 5 and 15 years

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
9.	Huang Y-K et al, 2007[E28]	1992-2005	9	Mean age 260 ± 215 days (20-790 days), oldest 31 years	Echo (9), Cath (8), LAM type A (3), A1/A2 (5), C (1), ASD(4)	Surgery (9), total correction (8), palliative op (1), RA (6), LA (3)	Mortality (0), mean follow-up (months-52.1 ± 43.6) (17-139), reoperation (0) NYHA I (8), lost to follow-up (1)
10.	Ozyuksel A et al, 2015[E20]	2001-2013	15	Age median 14 months (1 month – 7 years), infants (11)	Echo, Cath (15), cor triatriatum, concomitant cardiac pathology (14), ASD (3) PAPVC (1), ASD + PAPVC (4), complete AVSD (2), VSD (1), PDA (1), DORV (1)	Surgery (15), RA (13)-all had concomitant CHD	HD (1, 8%), mean follow-up 64 months (1-125 months), LD (0), reoperation (0)
11.	Al Qethamy HO et al, 2006[E6]	1983-2002	20	Mean 20 months (1-132 months) Males, 11	Echo (20), cor triatriatum (20), associated cardiac anomalies(12)	Trans-septal (19), LA (1)	HD (0), mean follow-up 31 months (2-156 months), LD (0), asymptomatic (14), NYHA (6)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
12.	Marin-Garcia J et al, 1975[E18]		20	Age <1 year=8, 1-5 years=6, >5 years=6, 38, 41 years=2, males 12	Necropsy studies (16), surgery (4), types – Diaphragmatic (10), Hourglass (3), Tubular (3), associated cardiac anomalies(7)	Not Stated	NA
13.	Humpl T et al, 2008[E35]	1951-2004	82	Median age 8 months (1 day-16.1years), females, 43	1954-81; Cath (70%), 1982 onwards echo (30%), associated cardiac lesions (77%), non-cardiac lesions (35%), chromosomal (12%)	Surgery (57%, 70%)-resection of fibro muscular diaphragm, no operation (14, 17%), died prior to intervention (11, 13%)	Total death (19, 23%), HD (5), LD (1), post-operative survival 94% at 3 months, 88% at 1 year, 86% at 5 years

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
14.	Kazanci S Y et al, 2012[E43]	1963-2010	66	Age <30 days (6) <12 months (37), median 7.2 months (2 days-14.6 years), males (36)	CTS (65), CTD (1), Associated CHD 49 (75%)	Surgery – excision membrane, associated procedures (47, 72%), RA or LA	HD (2), LD (1)-8.2 months post op with extra cardiac complete Fontan, median follow-up 5.4 years (3 days-44. 4 years) pulmonary vein angioplasty (2), follow-up – median 5.4 years, 48 survivors-NYHA I (39), II (7), III (2), NYHA III (2)-pulmonary vein angioplasty, recurrent insignificant membrane-6 (9%)

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
15.	Saxena P et al, 2014[E44]	1960-2012	29	Median 19 years (1 day-73 years), <1 month (3)	Isolated (5), LAM A (9), A1/A2 (13, 2), C (1), associated cardiac anomalies (21, 84%), pulmonary vein stenosis, CTS (25), CTD (4)	Surgery-CPB, cardioplegia, RA (16), LA (9)	HD (0), LD (2), median follow-up 6.6 years, survival at 10 years-83%, NYHA I (18, 72%), II (4, 16%), 2 with concomitant repair of complex anomalies died 2 and 5 months post op, LD (1) – unknown, no recurrent obstruction
16.	Krasemann Z et al, 2007[24]	1992-2003	10	Age 0.8 months (30 days-54 years)	Isolated CTS (1), Associated CHD (9)	Surgery (10)	HD (10%), follow-up-not mentioned

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
17.	Hamdan R et al, 2010[E3]	2010	1	40 years female	Clinical: OS ASD repair at age 5 years, paroxysmal palpitations, worsening dyspnea-4 months Persistent atrial flutter despite amiodarone + 3D β-blockers 2D TEE atrial membrane-LA 3D-TEE crescent shaped membrane within LA, Doppler gradient 8mmHg bicuspid aortic valve, dilated CS-LSVC	Operation: Radiofrequency flutter ablation-cavotricuspid isthmus ablation-SRdischarge oral anticoagulant β-blockers	Medical management

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
18.	Chen Q et al, 1999[E2]	1999	3	48 years, 35 years, 36 years Males, 2 Female, 1	1. Dyspnea, palpitation CCF (AF induced) 1994-deteriorated CXR: massive cardiomegaly, ECG, atrial fibrillation, RAD TTE,TEE; Large atria, intra-atrial membrane, 2.Dyspnea-diag as distal cardiomyopathy with atrial fibrillation, loud P2 Echo-CT PAP: 92/52 mmHg 3. 6 years age-murmur, AVSD, MR, split S2, echo-partial AVSD, large LSVC	Operation (2 patients): Cardiomegaly, dilated RV, tense PA, moderate PAH, membrane-IA-opening 2mm, calcified 2nd membrane above the PFO between pulmonary veins-lower chamber fenestrated and calcified. Membrane excised 3rd patient: operation-partial AVSD, CT 1 cm hole, LSVCLA via unroofed CS, membrane excised, IA baffle	Post: doing well short-term follow-up.

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
19.	McManus BM et al, 1982[17]	1982	1	31 year male	Opiate addict, IE staph aureus Severe AR	Operation: 3 aortic cusps, destroyed cusp excised, ring abscess-neck caudal to junction of left and posterior cusps Porcine bio: 23mm Died-narcotic overdose on 75th day No AR Necropsy-accessory chamber larger than RA, compressed the LA, blind pouch Diagnosis: Residual infection caudal to the bio-prosthesis ring false aneurysm.	Necropsy study

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
20.	Zaidi SJ et al, 2017[21]	2017	1	9 years, female	DORV (Taussig-Bing), TGA, complex VSD with inlet extension, straddling AV valves, valvar PS, juxtaposed atrial appendage. BT shunt (5 months), BDG (17 months) Extracardiac Fontan-aortic homograft 19mm (29 months) 7 years age-PLE, Echo- no Fontan pathway Obstruction 9 years age-increasing LA size-CTS, TEE-fibromuscular membrane (5 x 8 mm) Orifice gradient mean 7mmHg Cath: increased PCWP 15 mmHg, RVEDP (4-5 mmHg) Fontan pr. 20 mmHg	Operation: Resection CT membrane, atrial septectomy, Fontan fenestration, orifices pulmonary veins intact	Follow-up: doing well

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
21.	Oaks TE et al, 1995[16]	1995	1	42 years female	Idiopathic cardiomyopathy, cardiac transplant, CVP 15 mmHg Cardiac transplant (aged 38 years) Postop: deteriorating hemodynamics CVP: 22 mmHg, PAP 40/30 mmHg TTE: Large echo dense mass-mid LA obstructing LV inflow, gradient across the membrane 15 mmHg	CPB: LA suture line was patulous without adherent thrombus. 1.5 cm excessive atrial tissue was resected along half of atrial circumference, post op TEE: Marked reduction membrane Postop size, non-obstructed laminar flow to the MV	Postoperative recovery smooth Follow-up: 12 months-doing well.

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
22.	McGuire LB et al, 2015[E1]	2015	1	19 years male	Recurrent hemoptysis for 14 months prior, Heart murmur, Loud S1, S2 CXR: Pulmonary congestion, dilated MPA Prominent Kerley B lines, Barium swallow induction, BA filled esophagus PA angio-CT – resection-membrane	Massive postoperative bleeding Left thoracotomy-bleed from MV ring Postmortem CPVC as large as LA, dilated PA, CPPVC to true LA-2opneings 6mm and 2mm	Necropsy study

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
23.	Ludomirsky A et al, 1990[E16]		1	65 years male	Dyspnea, palpitation from age 29 years, cath at 30 years age, increased PAP, increased RV, LR shunt at SVC level.	Operation: 4mm thin-walled vessel from posterosuperior aspect left pulmonary hilum draining left brachiocephalic vein-ligated at brachiocephalic vein junction, anastomosed to LA 32 years later-dyspnea, palpitation TEE (intraoperative)-CT, MR Operation: Resection CT, mitral valvuloplasty	Discharged 1 week later

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
24.	Kumar VK et al, 2019[E15]	2009-2019	14	Age mean 12.14±9.97 years (1 year-29 years) Famels, 8	SOB (12), palpitation (4), cyanosis (1), CCF (1) Isolated CT (2) associated CHD (14), ASD (10), PAVSD (2), VSD (1), PS (1), DORV (1) Preoperative missed diagnosis (3) diagnosis-intraoperatively Severe PAH (8)	Excision CT, PTFE patch repair associated CHD repaired	HD (0) Mean follow=up: 58.06±30.73 months (20-120 months) Asymptomatic – no reoperation
25.	Jacobs A et al, 2006[E21]	2006	2	Case 1: 51 years male, Case 2: 79 years male	Case 1: unrepaired ASD- diag of PH, EchoCT, mild MR 3D echo-CT with communication between 2 chambers, OS-ASD Case 2: Cx spine # perioperative treatment, Echo-LVH mod CT 3D echo CT, large gap in membrane	Surgery – not done	NA

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
26.	Guvenc TS et al, 2012[E45]		2	31 years female, 27 years male	1.Pregnancy-SOB-CXR (N) 2D echo-CT with a large fenestration anteriorly 2.Chest pain x 1 month, 2D, 3D echo, CT incomplete membrane, ant opening all PV post chamber Both pressure gradient minimal Treadmill-Minimal symptoms	Surgery: not done	NA
27.	deBelder MA et al, 1992[E13]	1992	1	22 years male	SOB with Sydenham's chorea, S1-loud-diagnosis of MS, TTE-CTS TEE CT with a small defect, calcifies margin, PV-upper chamber	T- LA approach-excision of the membrane	Postop- 3 months-symptom free Nil medications.

S.No.	Authors, years	Period of study	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	Results/Follo up
28.	Sakamoto I etal, 1994[E30]	1994	2	25 years male, 26 years male	Both patients-No clinical findings, Normal heart sounds, TTE-anomalous membrane in LA, ASD (1), Spin-echo & Cine-MR-membrane in LA, Pulmonary veins connected to the accessory chamber, LAA to the true chamber	Anomalous membrane, 1 & 1.5cm fenestration confirmed on table Resection of the membrane	Not mentioned

Abbreviations: AF-atrial fibrillation, AR-aortic regurgitation, ASD-atrial septal defect, AV valves- atrioventricular valves, AVSD-atrio ventricular septal defect, BDG-bidirectional Glenn, Cath-catheterisation, CCF-congestive cardiac failure, CHD-congenital heart disease, CPB-cardiopulmonary bypass, CPVC-common pulmonary venous chamber, CS-coronary sinus, CT-cor triatriatum, CTD-cor triatriatum dexter, CTS-cor triatriatum sinister, CXR-chest X-ray, DHCA-deep hypothermic circulatory arrest, DORV-double outlet right ventricle, HD-hospital death, LAA-left atrial appendage, LA-left atrium, LD-late death, LSVC-left superior caval vein, LVH-left ventricular hypertrophy, LV-left ventricle, MR-mitral regurgitation, MV-mitral valve, OS-ASD-ostium secundum atrial septal defect, PAH-pulmonary artery hypertension, PAP-pulmonary artery pressure, PAPVC-partially anomalous pulmonary venous connection, PC-pulmonary chamber, PCWP-pulmonary capillary wedge pressure, PDA-patent ductus arteriosus, PND-paroxysmal nocturnal dyspnoea, Pre op-preoperative, PS-pulmonary stenosis, PV-pulmonary vein, RAD-right axis deviation, RA-right atrium, RVEDP-right ventricular end diastolic pressure, SR-sinus rhythm, SVC-superior vena cava, TEE-transesophageal echocardiography, TTE-transthoracic echocardiography, VC-vestibular chamber, VSD-ventricular septal defect

Table E2: Summary of the published investigations documenting the diagnosis of divided right atrium with or without associated congenital cardiac anomalies and its management

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
1.	Gerlis and Anderson, 1976[E112]	1976	1976	1	1	NA	No clinical history available. Necropsy-probable age between 3 and 6 months	No clinical history available. Necropsy-probable age between 3 and 6 months	No clinical history available. Necropsy-probable age between 3 and 6 months	No clinical history available. Necropsy-probable age between 3 and 6 months	Necropsy- fibromuscular partition within RA, OS ASD- 0.8 cm in diameter. Ebstein's anomaly (Imperfora	N f n l p t w E C A 0 i a E s a

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
2.	Alboliras ET et al, 1987[E130]	1987	1987	1	1	2.61 kg girl born to a 30 year old mother	ECG- congenital heat block 2D echo- fibromuscular large membrane dividing the RA. Additional findings- 5-8 mm VSD, aneurysmal membranous septum Pulmonary valve- thickened and domed.	ECG- congenital heat block 2D echo- fibromuscular large membrane dividing the RA. Additional findings- 5-8 mm VSD, aneurysmal membranous septum Pulmonary valve- thickened and domed.	ECG- congenital heat block 2D echo- fibromuscular large membrane dividing the RA. Additional findings- 5-8 mm VSD, aneurysmal membranous septum Pulmonary valve- thickened and domed.	ECG- congenital heat block 2D echo- fibromuscular large membrane dividing the RA. Additional findings- 5-8 mm VSD, aneurysmal membranous septum Pulmonary valve- thickened and domed.	Died- 2 days later- No surgery Postmorte A large septating membrane in the RA consistent with CTD Additional 4 small VSD's, PS	D d l N s F A s i n b i F c t w C A t 4 V F

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
3	Rao S et al, 2018[E150]	2018	2018	1	1	10 months old male	Persistent hypoxemia, O2 requirement– 1.5 lit throughout the day Foetal diagnosis- large ASD, PS, hypoplastic RV Comorbidities- bilateral chronic subdural hematomas post ventriculo-peritoneal shunt placement 4 months age- balloon pulmonary valvuloplasty Cyanosed- SaO_2- 80% TTE- Complex, mobile membranous struc-	Persistent hypoxemia, O2 requirement– 1.5 lit throughout the day Foetal diagnosis- large ASD, PS, hypoplastic RV Comorbidities- bilateral chronic subdural hematomas post ventriculo-peritoneal shunt placement 4 months age- balloon pulmonary valvuloplasty Cyanosed- SaO_2- 80% TTE- Complex, mobile membranous struc-	Persistent hypoxemia, O2 requirement– 1.5 lit throughout the day Foetal diagnosis- large ASD, PS, hypoplastic RV Comorbidities- bilateral chronic subdural hematomas post ventriculo-peritoneal shunt placement 4 months age- balloon pulmonary valvuloplasty Cyanosed- SaO_2- 80% TTE- Complex, mobile membranous struc-	Persistent hypoxemia, O2 requirement– 1.5 lit throughout the day Foetal diagnosis- large ASD, PS, hypoplastic RV Comorbidities- bilateral chronic subdural hematomas post ventriculo-peritoneal shunt placement 4 months age- balloon pulmonary valvuloplasty Cyanosed- SaO_2- 80% TTE- Complex, mobile membranous struc-	Biventricular repair	R r

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
4.	Aliyu I, Obrahim ZF, 2018[E120]	2018	2018	1	1	Age 3 months female	Central cyanosis (neonatal), tachypnoeic, tachycardic Apex beat-displaced SaO_2 85%-87% in room air CXR-cardiomegaly, increased pulmonary vascularity, ECG-biventricular hypertrophy TTE-thickened partition-RA, ASD, VSD (5 & 6 mm) CTD with TGA	Central cyanosis (neonatal), tachypnoeic, tachycardic Apex beat-displaced SaO_2 85%-87% in room air CXR-cardiomegaly, increased pulmonary vascularity, ECG-biventricular hypertrophy TTE-thickened partition-RA, ASD, VSD (5 & 6 mm) CTD with TGA	Central cyanosis (neonatal), tachypnoeic, tachycardic Apex beat-displaced SaO_2 85%-87% in room air CXR-cardiomegaly, increased pulmonary vascularity, ECG-biventricular hypertrophy TTE-thickened partition-RA, ASD, VSD (5 & 6 mm) CTD with TGA	Central cyanosis (neonatal), tachypnoeic, tachycardic Apex beat-displaced SaO_2 85%-87% in room air CXR-cardiomegaly, increased pulmonary vascularity, ECG-biventricular hypertrophy TTE-thickened partition-RA, ASD, VSD (5 & 6 mm) CTD with TGA	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
5.	Theodoropoulo KC et al, 2018[E151]	2018	2018	1	1	54 years old male	H/o lymphocytic hypophysitis and hypopituitarism- cerebellar infarcts on brain MRI TTE- Inj agitated saline– Intermittent clusters of bubbles in the non-opacified compartment with valsalva– appearance of a few bubbles into the left heart through ASD TEE and MRI-	H/o lymphocytic hypophysitis and hypopituitarism- cerebellar infarcts on brain MRI TTE- Inj agitated saline– Intermittent clusters of bubbles in the non-opacified compartment with valsalva– appearance of a few bubbles into the left heart through ASD TEE and MRI-	H/o lymphocytic hypophysitis and hypopituitarism- cerebellar infarcts on brain MRI TTE- Inj agitated saline– Intermittent clusters of bubbles in the non-opacified compartment with valsalva– appearance of a few bubbles into the left heart through ASD TEE and MRI-	H/o lymphocytic hypophysitis and hypopituitarism- cerebellar infarcts on brain MRI TTE- Inj agitated saline– Intermittent clusters of bubbles in the non-opacified compartment with valsalva– appearance of a few bubbles into the left heart through ASD TEE and MRI-	Surgery- not mentioned	S n m

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
6.	Gussenhoven WJ et al, 1982 E113	1982	1982	1	1	13 years old female	Progressively limited exercise tolerance, mild cyanosis Gr 4/6 mid-systolic murmur- 2^{nd} ICS, split S2 Echo M-mode – multiple blurred echoes in diastole posterior to the ATL. Echoes also in the RVOT 2D-echo one or probably 2 soft, thin walled structures originating in the RA.	Progressively limited exercise tolerance, mild cyanosis Gr 4/6 mid-systolic murmur- 2^{nd} ICS, split S2 Echo M-mode – multiple blurred echoes in diastole posterior to the ATL. Echoes also in the RVOT 2D-echo one or probably 2 soft, thin walled structures originating in the RA.	Progressively limited exercise tolerance, mild cyanosis Gr 4/6 mid-systolic murmur- 2^{nd} ICS, split S2 Echo M-mode – multiple blurred echoes in diastole posterior to the ATL. Echoes also in the RVOT 2D-echo one or probably 2 soft, thin walled structures originating in the RA.	Progressively limited exercise tolerance, mild cyanosis Gr 4/6 mid-systolic murmur- 2^{nd} ICS, split S2 Echo M-mode – multiple blurred echoes in diastole posterior to the ATL. Echoes also in the RVOT 2D-echo one or probably 2 soft, thin walled structures originating in the RA.	Surgery- Large membrane representing a persistent right sinus venosus valve was found and resected. Pulmonary valve normal. White finger-like membrane 7 x 3 cm, broad-stalked cut edges, fenestrated	[illegible]

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
7.	Hurtado-Sierra D et al, 2020[E124]	2020	2020	1	1	18 days–Term new born	Intermittent cyanosis, no respiratory distress, O_2 saturation 65% CXR-Normal, SaO_2 85% TTE- a large undulating membrane dividing the RA into two chambers Agitated saline-accumulation of microbubbles in the postero-medial chamber	Intermittent cyanosis, no respiratory distress, O_2 saturation 65% CXR-Normal, SaO_2 85% TTE- a large undulating membrane dividing the RA into two chambers Agitated saline-accumulation of microbubbles in the postero-medial chamber	Intermittent cyanosis, no respiratory distress, O_2 saturation 65% CXR-Normal, SaO_2 85% TTE- a large undulating membrane dividing the RA into two chambers Agitated saline-accumulation of microbubbles in the postero-medial chamber	Intermittent cyanosis, no respiratory distress, O_2 saturation 65% CXR-Normal, SaO_2 85% TTE- a large undulating membrane dividing the RA into two chambers Agitated saline-accumulation of microbubbles in the postero-medial chamber	Surgery-Not mentioned	S N m

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
8.	Yamaguchi R et al, 2013[E127]	2013	2013	1	1	82 years Female	Palpitation, chest oppression, SOB CXR: CTR 64%, ECG-Atrial fib 92/min SaO_2– 96%, grade IV/VI systolic murmur TTE-dilated right heart Volume overload, LVEF normal TV morphology-normal Systolic gradient across TV 46 mmHg Large band in the RA dividing into two chambers, no	Palpitation, chest oppression, SOB CXR: CTR 64%, ECG-Atrial fib 92/min SaO_2– 96%, grade IV/VI systolic murmur TTE-dilated right heart Volume overload, LVEF normal TV morphology-normal Systolic gradient across TV 46 mmHg Large band in the RA dividing into two chambers, no	Palpitation, chest oppression, SOB CXR: CTR 64%, ECG-Atrial fib 92/min SaO_2– 96%, grade IV/VI systolic murmur TTE-dilated right heart Volume overload, LVEF normal TV morphology-normal Systolic gradient across TV 46 mmHg Large band in the RA dividing into two chambers, no	Palpitation, chest oppression, SOB CXR: CTR 64%, ECG-Atrial fib 92/min SaO_2– 96%, grade IV/VI systolic murmur TTE-dilated right heart Volume overload, LVEF normal TV morphology-normal Systolic gradient across TV 46 mmHg Large band in the RA dividing into two chambers, no	Treatment-Medical-diuretics, vasodilators, antiplatelets	T M d i v s t a p

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
9.	Mackman CA et al, 2015[E161]	2000-2013	2000-2013	3	3	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	**Case 1:** 3.2 kg term female, intermittent SaO_2 70-80% Echo-VSD, PFO Membrane RA-CTD RL across PFO, elevated RAP Surgery-resection-membrane+ASD closure Home-6th postoperative day, No O_2 requirement, no complications **Case 2:** 3.2 kg female SaO_2 73%, physical examination-normal Echo-	

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
10.	Low TT et al, 2013[E162]	2013	2013	1	1	50 years old male	Atrial flutter CHF, prior CABG – (2002) TTE dilated LV, eccentric LVH, mild MR, TR TEE- to rule out intracardiac thrombus prior to radiofrequency ablation+CTD 3D echo-CTD with LAA thrombus	Atrial flutter CHF, prior CABG – (2002) TTE dilated LV, eccentric LVH, mild MR, TR TEE- to rule out intracardiac thrombus prior to radiofrequency ablation+CTD 3D echo-CTD with LAA thrombus	Atrial flutter CHF, prior CABG – (2002) TTE dilated LV, eccentric LVH, mild MR, TR TEE- to rule out intracardiac thrombus prior to radiofrequency ablation+CTD 3D echo-CTD with LAA thrombus	Atrial flutter CHF, prior CABG – (2002) TTE dilated LV, eccentric LVH, mild MR, TR TEE- to rule out intracardiac thrombus prior to radiofrequency ablation+CTD 3D echo-CTD with LAA thrombus	Treatment oral anticoagulation + atrial flutter ablation	T o a a t a f a

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
11.	Vukovic PM et al, 2014[E163]	2014	2014	1	1	43 years old female	SOB, NYHA-III, no signs of CHF TTE-2.5 x 3.5 cm ASD, QP:QS= 3:1 TEE-CTD Treatment: Percutaneous balloon dilation disrupted the membrane, enlarged the communication complete absence of the inferior rim-ASD no tissue was available for device	SOB, NYHA-III, no signs of CHF TTE-2.5 x 3.5 cm ASD, QP:QS= 3:1 TEE-CTD Treatment: Percutaneous balloon dilation disrupted the membrane, enlarged the communication complete absence of the inferior rim-ASD no tissue was available for device	SOB, NYHA-III, no signs of CHF TTE-2.5 x 3.5 cm ASD, QP:QS= 3:1 TEE-CTD Treatment: Percutaneous balloon dilation disrupted the membrane, enlarged the communication complete absence of the inferior rim-ASD no tissue was available for device	SOB, NYHA-III, no signs of CHF TTE-2.5 x 3.5 cm ASD, QP:QS= 3:1 TEE-CTD Treatment: Percutaneous balloon dilation disrupted the membrane, enlarged the communication complete absence of the inferior rim-ASD no tissue was available for device	Surgical closure: resection of the membrane closure-ASD 3 x 2.5 cm pericardial patch	[illegible]

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery
12.	Omeje B et al, 2015[E140]	2015	2015	1	1	7 years old female	Type I neurofibromatosis Echo-hypoplastic abdominal aorta, CoA CTD with a 5.7 cm opening 7.8 mmHg pressure gradient between atrial chambers MRI: Long segment CoA, proximal DTA, post. stenotic dilatation, CTD Treatment: Stent-CoA segment 34 mm	Type I neurofibromatosis Echo-hypoplastic abdominal aorta, CoA CTD with a 5.7 cm opening 7.8 mmHg pressure gradient between atrial chambers MRI: Long segment CoA, proximal DTA, post. stenotic dilatation, CTD Treatment: Stent-CoA segment 34 mm	Type I neurofibromatosis Echo-hypoplastic abdominal aorta, CoA CTD with a 5.7 cm opening 7.8 mmHg pressure gradient between atrial chambers MRI: Long segment CoA, proximal DTA, post. stenotic dilatation, CTD Treatment: Stent-CoA segment 34 mm	Type I neurofibromatosis Echo-hypoplastic abdominal aorta, CoA CTD with a 5.7 cm opening 7.8 mmHg pressure gradient between atrial chambers MRI: Long segment CoA, proximal DTA, post. stenotic dilatation, CTD Treatment: Stent-CoA segment 34 mm	Surgery: CTD with a 6 mm opening in the fibrous tissue Intermittent circulatory arrest for access to IVC-extensive resection of the fibromuscular tissue More than 30^{th} postoperative day-pericardial + right pleural effusion pericardial drainage (low CO) ECMO x 1 wk

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
13.	Haboub and Drighil A, 2019[E145]	2019	2019	1	1	3 years old male	SOB, parasternal lift, grade 4/6 ESM, SaO_2 99% on room air Echo-discrete circumferential subpulmonic membrane and right atrial membrane RV-PA gradient 85 mmHg Treatment: Balloon dilation subpulmonic membrane, gradient 85 mmHg 50 mmHg Follow-up: TTE, resid-	SOB, parasternal lift, grade 4/6 ESM, SaO_2 99% on room air Echo-discrete circumferential subpulmonic membrane and right atrial membrane RV-PA gradient 85 mmHg Treatment: Balloon dilation subpulmonic membrane, gradient 85 mmHg 50 mmHg Follow-up: TTE, resid-	SOB, parasternal lift, grade 4/6 ESM, SaO_2 99% on room air Echo-discrete circumferential subpulmonic membrane and right atrial membrane RV-PA gradient 85 mmHg Treatment: Balloon dilation subpulmonic membrane, gradient 85 mmHg 50 mmHg Follow-up: TTE, resid-	SOB, parasternal lift, grade 4/6 ESM, SaO_2 99% on room air Echo-discrete circumferential subpulmonic membrane and right atrial membrane RV-PA gradient 85 mmHg Treatment: Balloon dilation subpulmonic membrane, gradient 85 mmHg 50 mmHg Follow-up: TTE, resid-	Surgery: Planned died of appendicular perforation prior to surgery	S F d a d p r p t s

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
14.	Jones RN and Niles NB 1968[E115]	1968	1968	1	1	10 years old male	Increasing cyanosis at 2 months age Diagnosed as tricuspid atresia with interatrial and possibly VSD Potts aorticopulmonary shunt-age 4½ months Cyanosis-decreased, CCF improved, subnormal somatic growth Recurrent syncope-cath-inadequate shunt flow, RAP-raised Sail like spinna-	Increasing cyanosis at 2 months age Diagnosed as tricuspid atresia with interatrial and possibly VSD Potts aorticopulmonary shunt-age 4½ months Cyanosis-decreased, CCF improved, subnormal somatic growth Recurrent syncope-cath-inadequate shunt flow, RAP-raised Sail like spinna-	Increasing cyanosis at 2 months age Diagnosed as tricuspid atresia with interatrial and possibly VSD Potts aorticopulmonary shunt-age 4½ months Cyanosis-decreased, CCF improved, subnormal somatic growth Recurrent syncope-cath-inadequate shunt flow, RAP-raised Sail like spinna-	Increasing cyanosis at 2 months age Diagnosed as tricuspid atresia with interatrial and possibly VSD Potts aorticopulmonary shunt-age 4½ months Cyanosis-decreased, CCF improved, subnormal somatic growth Recurrent syncope-cath-inadequate shunt flow, RAP-raised Sail like spinna-	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
15.	Verel D et al, 1970[E101]	1970	1970	1	1	21 years female	1st diag- Ebstein's anomaly – 2 years back, no cyanosis/clubbing CXR: Atrial shadow posteriorly enlarged Cath- Normal intracardiac pressure, SaO_2 96% Flask-shaped chamber behind the main chamber of RA; IVC-communicating with the lower extremity posterior chamber-diag	1st diag- Ebstein's anomaly – 2 years back, no cyanosis/clubbing CXR: Atrial shadow posteriorly enlarged Cath- Normal intracardiac pressure, SaO_2 96% Flask-shaped chamber behind the main chamber of RA; IVC-communicating with the lower extremity posterior chamber-diag	1st diag- Ebstein's anomaly – 2 years back, no cyanosis/clubbing CXR: Atrial shadow posteriorly enlarged Cath- Normal intracardiac pressure, SaO_2 96% Flask-shaped chamber behind the main chamber of RA; IVC-communicating with the lower extremity posterior chamber-diag	1st diag- Ebstein's anomaly – 2 years back, no cyanosis/clubbing CXR: Atrial shadow posteriorly enlarged Cath- Normal intracardiac pressure, SaO_2 96% Flask-shaped chamber behind the main chamber of RA; IVC-communicating with the lower extremity posterior chamber-diag	Surgery: Not mentioned	S N m

S. No.	**Authors, years**	**Period of study**	**Period of study**	**No. of patients**	**No. of patients**	**Age, sex**	**Anatomy / Coexisting lesions / Symptoms / Investigations**	**Anatomy / Coexisting lesions / Symptoms / Investigations**	**Anatomy / Coexisting lesions / Symptoms / Investigations**	**Anatomy / Coexisting lesions / Symptoms / Investigations**	**Surgery**	**S**
16.	Hansing CE et al 1972[E102]	1972	1972	1	1	25 years old male	Intermittent cyanosis at birth SOB- 5 years, cyanosis, clubbing Cath (21 years age)- diagnosed ASD Recurrent syncope, progressive SOB Recath- ASD, RA-RV gradient 3 mm Hg. Cineangiogram- RA membrane above the tricuspid valve	Intermittent cyanosis at birth SOB- 5 years, cyanosis, clubbing Cath (21 years age)- diagnosed ASD Recurrent syncope, progressive SOB Recath- ASD, RA-RV gradient 3 mm Hg. Cineangiogram- RA membrane above the tricuspid valve	Intermittent cyanosis at birth SOB- 5 years, cyanosis, clubbing Cath (21 years age)- diagnosed ASD Recurrent syncope, progressive SOB Recath- ASD, RA-RV gradient 3 mm Hg. Cineangiogram- RA membrane above the tricuspid valve	Intermittent cyanosis at birth SOB- 5 years, cyanosis, clubbing Cath (21 years age)- diagnosed ASD Recurrent syncope, progressive SOB Recath- ASD, RA-RV gradient 3 mm Hg. Cineangiogram- RA membrane above the tricuspid valve	Surgery- RA membrane, Tricuspid valve- 25 mm diameter, small ASD Resection of RA membrane + PFO closed	S F n b T p v 2 c t s A F t F n b - c

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
17.	Goldfarb A et al, 1989[E106]	1989	1989	1	1	30 years old female	TT Echo-mitral valve prolapse with left atrial membrane causing partition TEE-Membrane both within the RA and LA MRI-Non-obstructive, incomplete, membranous partition both within RA and LA	TT Echo-mitral valve prolapse with left atrial membrane causing partition TEE-Membrane both within the RA and LA MRI-Non-obstructive, incomplete, membranous partition both within RA and LA	TT Echo-mitral valve prolapse with left atrial membrane causing partition TEE-Membrane both within the RA and LA MRI-Non-obstructive, incomplete, membranous partition both within RA and LA	TT Echo-mitral valve prolapse with left atrial membrane causing partition TEE-Membrane both within the RA and LA MRI-Non-obstructive, incomplete, membranous partition both within RA and LA	Surgery: Not performed	S N p

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
18.	Chiari H, 1897[E149]	1897	1897	1	1	$7\frac{1}{2}$ years female	Cyanosis since birth Additional anomalies-PA, HRV, HTV, PDA, PFO, ASD	Cyanosis since birth Additional anomalies-PA, HRV, HTV, PDA, PFO, ASD	Cyanosis since birth Additional anomalies-PA, HRV, HTV, PDA, PFO, ASD	Cyanosis since birth Additional anomalies-PA, HRV, HTV, PDA, PFO, ASD	Nil intervention	N
19.	Doucette J and Knoblich R, 1963[E103]	1963	1963	1	1	6 weeks male	Cyanosis at birth Cath-CTD, PA, HRV, HTV, PFO, PDA Died after surgery	Cyanosis at birth Cath-CTD, PA, HRV, HTV, PFO, PDA Died after surgery	Cyanosis at birth Cath-CTD, PA, HRV, HTV, PFO, PDA Died after surgery	Cyanosis at birth Cath-CTD, PA, HRV, HTV, PFO, PDA Died after surgery	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
20.	Kaufman SL, Anderson DH, 1963[E104]	1963	1963	1	1	2 days male	Cyanosis + respiratory distress from 12 hours age Additional anomalies, PA, HRV, HTV, PDA, ASP	Cyanosis + respiratory distress from 12 hours age Additional anomalies, PA, HRV, HTV, PDA, ASP	Cyanosis + respiratory distress from 12 hours age Additional anomalies, PA, HRV, HTV, PDA, ASP	Cyanosis + respiratory distress from 12 hours age Additional anomalies, PA, HRV, HTV, PDA, ASP	Surgery: Modified Potts procedure	S M f P p
21.	Dubin IN and Hollinshead WH, 1944[E153]	1944	1944	1	1	5 days female	Cyanosis from birth Diagnosis-CTD, additional anomalies Hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis from birth Diagnosis-CTD, additional anomalies Hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis from birth Diagnosis-CTD, additional anomalies Hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis from birth Diagnosis-CTD, additional anomalies Hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery
22.	Folger GM, 1968[E152]	1968	1968	2	2	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar	**Case 1:** 9 years female Cyanosis from 3 months of age Diagnosis: CTD, additional PA, HRV, HTV, PDA, ASD Surgery: Resection of the membrane, pulmonary and tricuspid valvotomy Died after surgery **Case 2:** 5½ years male Cyanosis since birth, SOB Diagnosis: CTD, supra-valvar

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23.	Runcie J et al, 1968[E154]	1968	1968	1	1	26 years female	Constrictive pericarditis Obstruction of SVC, IVC	Constrictive pericarditis Obstruction of SVC, IVC	Constrictive pericarditis Obstruction of SVC, IVC	Constrictive pericarditis Obstruction of SVC, IVC	Surgery: pericardiec	S
24.	Panhold J et al, 1937[E156]	1937	1937	1	1	2 days female	Cyanosis and respiratory distress from CCF Diagnosis: CTD, hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis and respiratory distress from CCF Diagnosis: CTD, hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis and respiratory distress from CCF Diagnosis: CTD, hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	Cyanosis and respiratory distress from CCF Diagnosis: CTD, hypoplastic pulmonary trunk, HTV, dilated RV, PFO, PDA	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
25.	Ruggieri A et al, 1935[E157]	1935	1935	1	1	42 years female	Cyanosis, syncope and SOB since age 12 years, CNS disease since age 38 Diagnosis: CTD, small pulmonary trunk, PFO, ASD	Cyanosis, syncope and SOB since age 12 years, CNS disease since age 38 Diagnosis: CTD, small pulmonary trunk, PFO, ASD	Cyanosis, syncope and SOB since age 12 years, CNS disease since age 38 Diagnosis: CTD, small pulmonary trunk, PFO, ASD	Cyanosis, syncope and SOB since age 12 years, CNS disease since age 38 Diagnosis: CTD, small pulmonary trunk, PFO, ASD	NA	N
26.	Kettler L et al, 1934[E158]	1934	1934	1	1	60 years male	Asymptomatic Diagnosis: CTD, wide pulmonary trunk, RA dilation, PFO, ASD	Asymptomatic Diagnosis: CTD, wide pulmonary trunk, RA dilation, PFO, ASD	Asymptomatic Diagnosis: CTD, wide pulmonary trunk, RA dilation, PFO, ASD	Asymptomatic Diagnosis: CTD, wide pulmonary trunk, RA dilation, PFO, ASD	NA	N

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
27.	Sternberg C et al, 1913[E160]	1913	1913	1	1	21 years female	Asymptomatic. Diagnosis: CTD, PFO, ASD	Asymptomatic. Diagnosis: CTD, PFO, ASD	Asymptomatic. Diagnosis: CTD, PFO, ASD	Asymptomatic. Diagnosis: CTD, PFO, ASD	NA	Nonc
28.	Gombert H et al, 1933[E159]	1933	1933	1	1	42 years female	Asymptomatic. Diagnosis: CTD, small PA, HRV, PFO, ASD	Asymptomatic. Diagnosis: CTD, small PA, HRV, PFO, ASD	Asymptomatic. Diagnosis: CTD, small PA, HRV, PFO, ASD	Asymptomatic. Diagnosis: CTD, small PA, HRV, PFO, ASD	NA	Nonc

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery
29.	Trento A ert al, 1988[E111]	1988	1988	Clinical: N=2	Clinical: N=2	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery	Case 1: 1-day-old infant, increasing cyanosis from birth, Echo-Right atrial partition obstructing the Tricuspid-orifice, Cath: mean RAP, mean IVC pressure= 2mmHg, RV 40/0 mmHg, spinnaker like structure into the RV, prominent Eustachian and thebesian valves, surgery

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	Necropsy: N=14	N
S. No.	**Atrial arrangement**	**Atrial arrangement**	**AV connection**	**AV connection**	**VA connection**	**VA connection**	**VA connection**	**Associated defects**	**Chiari network**	**Persistent venous valve**	**Persistent venous valve**	**C**
1	Usual	Usual	Concordant	Concordant	Concordant	Concordant	Concordant	TOF	+	-	-	M r
2	Usual	Usual	Concordant	Concordant	Discordant	Discordant	Discordant	Complete TGA	+	-	-	P t u c s c p e s
3	Usual	Usual	Concordant	Concordant	Discordant	Discordant	Discordant	Complete TGA	+	-	-	P t c c T c t C
4	Usual	Usual	Concordant	Concordant	Common trunk	Common trunk	Common trunk	Interrupted aorta, VSD with muscle rim	+	-	-	V f r w t u c s c p e s

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
5	Usual	Usual	Concordant	Concordant	Concordant	Concordant	Concordant	Oval fossa defect	+	-	-	M n
6	Usual	Usual	Absent RAVC	Absent RAVC	Discordant	Discordant	Discordant	Oval fossa defect	+	-	-	M n
7	Usual	Usual	Concordant	Concordant	Aortic atresia	Aortic atresia	Aortic atresia	Imperforate LAVV	+	+	+	P e c i t S I C
8	Usual	Usual	Concordant	Concordant	Concordant	Concordant	Concordant	TOF	-	+	+	P e c i t S I C
9	Usual	Usual	Concordant	Concordant	Pulmonary atresia	Pulmonary atresia	Pulmonary atresia	Ebstein's malformation, IVS	-	+	+	P a v n c r t e t t s t s

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
10	Usual	Usual	Absent RAVC	Absent RAVC	Concordant	Concordant	Concordant	Nonrestrictive VSD	-	+	+	P a v r s
11	Usual	Usual	Absent RAVC	Absent RAVC	Concordant	Concordant	Concordant	Left atrium to morphologic RV	-	+	+	P a v r c r t e t t s t s
12	Usual	Usual	Absent RAVC	Absent RAVC	Concordant	Concordant	Concordant	Restrictive VSD	-	+	+	P S I a s c r s r a r p

S. No.	Authors, years	Period of study	Period of study	No. of patients	No. of patients	Age, sex	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Anatomy / Coexisting lesions / Symptoms / Investigations	Surgery	S
13	Usual	Usual	Absent RAVC	Absent RAVC	Concordant	Concordant	Concordant	Atresia with rudimentary RV	-	+	+	P a v r s r e t t s t s
14	Usual	Usual	Double inlet LV	Double inlet LV	Concordant	Concordant	Concordant	Ebstein's malformation	-	+	+	P a v r s r e t t s t s

Abbreviations: ATL-anterior tricuspid leaflet, CABG-coronary artery bypass grafting, CS- coronary sinus, CCF-congestive cardiac failure, CFB-central fibrous body, CHF-congestive heart failure, CNS-central nervous system, COA-coarctation of the aorta, CTD-cor triatriatum dexter, CTR-cardiothoracic ratio, CXR-chest x-ray, DTA-descending thoracic aorta, ECG-electrocardiogram, ECMO-extra corporeal membrane oxygenation, ESM-ejection systolic murmur, HRV-hypoplastic right ventricle, HTV-hypoplastic tricuspid valve, ICS-inter costal space, ICV-inferior caval vein, IVC-inferior vena cava, IXCV-inferior caval vein, LAA-left atrial appendage, LA-left atrium, LAVV- left atrio ventricular valve, L–R-left-to-right shunt, LVEF-left ventricular ejection fraction, LVH-left ventricular hypertrophy, LV-left ventricle, MRI-magnetic resonance imaging, MR-mitral regurgitation, NA-not available, NYHA-New York Heart Association, OS ASD-ostium Secundum Atrial septal defect, PA-pulmonary artery, PDA-persistent ductus arteriosis, PDA-persistent ductus arteriosus, PFO-patent foramen ovale, PFO-persistent foramen ovale, PGE-1-prostaglandin E1, PS-pulmonary stenosis, PS-pulmonic stenosis, QP:QS-systemic-to-pulmonary blood flow, RAP-right atrial pressure, RAP-right atrial pressure, RA-right atrium, RAVC- right atrio ventricular connection, RVEDV-right ventricular

end-diastolic volume, RVEF-right ventricular ejection fraction, RVOT-right ventricular outflow tract, RV-right ventricle, SaO_2-systemic arterial oxygen saturation, SOB-shortness of breath, TEE-transesophageal echocardiography, TGA-transposition of the great arteries, TR-tricuspid regurgitation, TR-tricuspid regurgitation, TTE-transthoracic Echocardiography, TV-tricuspid valve, VSD-ventricular Septal defect