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# Surgical repair of atrioventricular septal defect with common atrioventricular junction when associated with tetralogy of Fallot or double outlet right ventricle

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The association of atrioventricular septal defect with common atrioventricular junction and malformations of the ventricular outflow tracts presents a significant challenge for the surgeon. In the most common of these, the association with tetralogy of Fallot, several surgical techniques have been described, and shown to deliver excellent results.[1–10] On the other hand, in the setting of more extreme malformations, such as double-outlet right ventricle, discordant ventriculo-arterial connections, or common arterial trunk, albeit rare lesions, the combination presents a more formidable surgical challenge, as evidenced by the few reports of successful repair of these lesions. This challenge is both physiological, when dealing with a very sick neonate or infant, as well as anatomical in terms of the complexity of the malformation and the ability to achieve a successful biventricular repair. Our goal in this review is to discuss the surgical treatment in the setting of tetralogy of Fallot and double outlet right ventricle, with emphasis on biventricular repair.

## Incidence and classification

Although the incidence of common atrioventricular junction in the setting of tetralogy of Fallot can be estimated from published clinical series, the true incidence of more complex anomalies, such as double outlet right ventricle, is harder to gauge due to the paucity of published reports. The true incidence of these anomalies can be obtained from a landmark paper published by Bharati and associates,[11] which described the association of a common atrioventricular junction with tetralogy of Fallot, double outlet right ventricle, or discordant ventriculo-arterial connections. These anomalies were found in 81 out of 507 specimens with common atrioventricular canal, with 30 specimens having tetralogy of Fallot, 34 double outlet right ventricle, and 17 discordant ventriculo-arterial connections. Bharati et al.[11] further found that, amongst the cases with tetralogy of Fallot, almost all of the specimens had a common atrioventricular valvar orifice, which they nominated as "complete" defects, the remainder having a common atrioventricular junction, but with separate valvar orifices for the two ventricles, albeit, with the potential for interventricular shunting. They called these "intermediate" defects, although the junction is just as common, and the valve has the same basic morphology, apart from the separateness of the valvar orifices for the right and left ventricles. Similar proportions of morphologies were observed among the cases with double outlet right ventricle and transposition. It is well recognized that, in these settings, there can be any arrangement of the atrial appendages, common atrioventricular junction being frequently found in the setting of heterotaxy syndromes.

The classification of the specific morphology of the common atrioventricular junction in the setting of tetralogy of Fallot or transposition is straightforward. On the other hand, the classification with double outlet from the right ventricle can be more complex and

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controversial. Some have suggested that, when both arterial trunks arise from the right ventricle, the ventricular component of the atrioventricular septal defect always has a subaortic extension. These cases would obviously be along a continuum with tetralogy of Fallot, albeit with more extreme aortic override. The majority of the published cases of surgical repair of double outlet right ventricle with a common atrioventricular junction are certainly of this type. In the large autopsy series collected by Bharati et al.,[11] nonetheless, it is clear that the anatomic types of double outlet right ventricle are similar to those seen in patients with separate right and left atrioventricular junctions. Among the 34 specimens collected by them with double outlet right ventricle and common atrioventricular junction, all the expected positions for the interventricular communication were observed. In 15 specimens, the interventricular communication had a subaortic extension, as in the majority of the published clinical cases. In another 15 specimens, however, the interventricular communication was restricted to the inlet of the right ventricle, with no subarterial extension and was therefore of the non-committed type. In 3 specimens, the defect was doubly committed, while subpulmonary extension was seen in the final specimen. If replicated in clinical practice, which seems likely, these findings have great surgical significance, as they obviously would influence the types of surgical repair.

If the interventricular communication has a subaortic extension, then the differentiation between tetralogy of Fallot and double outlet right ventricle may also be difficult and controversial, the same problems existing in the setting of separate atrioventricular junctions. This difficulty in classification is highlighted in a report by Karl et al.,[6] who grouped these patients together, using the terms interchangeably. On the other hand, when interventricular communication has no subarterial extension, it is of necessity non-committed. The aorta then arises completely from the right ventricle along with the pulmonary trunk, and it is then clear that we are dealing with common atrioventricular junction and unequivocal double outlet right ventricle.

## Repair of common atrioventricular junction in the setting of tetralogy of Fallot

Surgical repair of tetralogy of Fallot with common atrioventricular junction presents several challenges and controversies. These include the role of initial palliation and delayed repair as opposed to early primary repair, the use of a single patch as opposed to two patches, and transventricular in contrast to transatrial repair. The timing of repair is to some extent dictated by the degree of cyanosis, which is determined by the degree of obstruction of the right ventricular outflow tract. Congestive heart failure is infrequent because of the restricted flow of blood to the lungs. Although a significant number of patients have been initially palliated by construction of a systemic-to-pulmonary shunt in most of the published series, some authors have recently advocated early primary repair.[7,8]

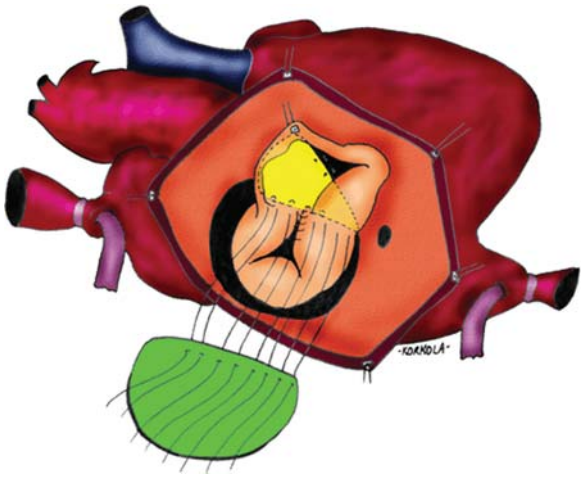


**Figure 1.**
*The cartoon shows how the common atrioventricular junction can be septated using two patched when combined with tetralogy of Fallot. Note that the ventricular patch is shaped like a comma.*

In addition to the goals of repair of the atrioventricular septal defect, which are to achieve as competent as possible left and right atrioventricular valves and close the interatrial and interventricular communications, the added presence of the tetralogy mandates the avoidance of both subaortic and subpulmonary obstruction subsequent to the repair. The common atrioventricular junction can be septated either using two patches[3,5,6,9,10] or a single patch.[7,12] The presence of tetralogy of Fallot imposes some modifications in these techniques, in order to avoid residual subaortic obstruction. When using two patches (Fig. 1), the ventricular patch needs to be wider near the subaortic area to allow enough redundancy at the anterosuperior end of the interventricular communication, and thus avoid obstruction of the left ventricular outflow tract.[12] The ventricular patch, therefore, needs to be shaped like a comma.[6] If a single patch is used (Fig. 2), the superior bridging leaflet is divided obliquely to the right, paralleling the malaligned outlet septum, in order to prevent subaortic stenosis.[7] The obstructed right ventricular outflow tract is relieved by techniques determined by the degree and the level of the obstruction. All efforts should be made to preserve a functional pulmonary valve, unless it is dysplastic and the ventriculo-arterial junction severely hypoplastic.

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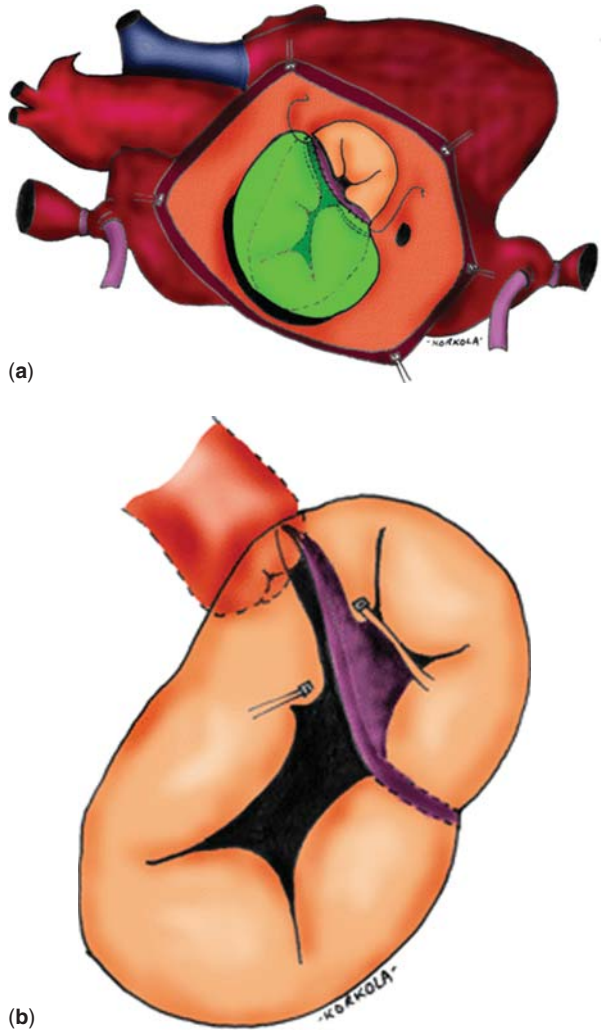


**Figure 2.**
*(**a**) shows the technique of using a single patch for correction of tetralogy of Fallot in the setting of a common atrioventricular junction. (**b**) shows how the ventricular component of the patch is deviated rightward to surround the overriding part of the aorta.*

If the pulmonary valve is preserved, the subpulmonary obstruction is repaired by incision and insertion of an autologous pericardial patch. If the pulmonary valve is dysplastic and the ventriculo-arterial junction is hypoplastic, a transjunctional patch may be needed. It may be prudent to place a valved conduit between the right ventricle and the pulmonary arteries if a less than perfect repair of the atrioventricular valves is anticipated, particularly the left valve, or when there is distal pulmonary arterial hypoplasia. Transatrial and transpulmonary repair has recently been reported with favourable outcome.[6,10] Most surgeons, nonetheless, advocate a transventricular approach when the common atrioventricular junction is accompanied by tetralogy of Fallot.

## Repair of atrioventricular septal defect with common atrioventricular junction and double outlet right ventricle

As discussed above, the association of common atrioventricular junction and double outlet right ventricle is well documented in autopsy series.[11,13–15] As emphasized in the previous section, the classification of this combination is best determined on the extent of subarterial extension, or lack of such extension, of the ventricular component of the atrioventricular septal defect.[11] This useful classification has important surgical implications as to the type of biventricular repair that will be feasible. Also of surgical importance is the presence or absence of ventricular hypoplasia if contemplating biventricular repair. In another autopsy study, Van Praagh et al.[13] found varying degrees of right or left ventricular hypoplasia in 35 hearts with double outlet right ventricle in the setting of a common atrioventricular junction.

Despite these reports of pathological specimens, accounts of successful surgical repair are less frequent.[1,6,16–20] Surgical techniques to deal with this complex association have been described, and depend mainly on the degree of extension of the ventricular component of the atrioventricular septal defect towards the great arteries.[1,16–19] When there is subaortic extension, the repair is similar to that used for tetralogy of Fallot and common atrioventricular junction,[6] using a patch shaped like a comma to direct blood from the left ventricle to the aorta,[1,16,17] accompanied of course by septation of the common atrioventricular junction. If the subpulmonary outflow tract is obstructed, it is repaired as described above. As pointed out by Karl et al.,[6] the greater the aortic override above the right ventricle, the larger will need to be the head of the comma if subaortic stenosis is to be avoided.[6]

If the ventricular component of the atrioventricular septal defect extends to the subpulmonary area, the common atrioventricular junction can be septated with a patch shaped like a comma so that the left ventricle is tunnelled to the pulmonary trunk, combining this with an arterial switch procedure.[18] Should the interventricular communication be non-committed, however, in other words does not extend toward the ventricular outflow tracts, then of necessity there will be a significant distance between the defect and the aorta (Fig. 3). In this setting, anatomic biventricular repair is extremely challenging, and as far as we are aware, has not been reported until recently. Pacifico et al.[16] have now described 2 patients with double outlet right ventricle and common atrioventricular junction in the setting of mirror-imaged arrangement, but without any subarterial extension of the interventricular communication. They septated the common atrioventricular junction by closing the atrial

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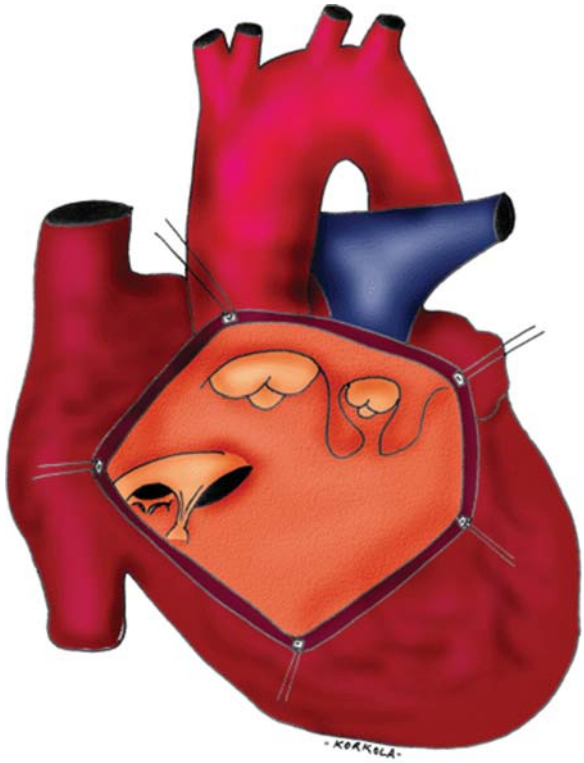

**Figure 3.**
*The cartoon shows the morphology of double outlet right ventricle in the setting of a common atrioventricular junction, with the ventricular component of the atrioventricular septal defect in non-committed position. In the illustrated heart, there is also subpulmonary stenosis.*

and ventricular components of the septal defect, closed also the pulmonary trunk, placed a conduit from the left ventricle to the pulmonary arteries, and performed an atrial switch operation.[16] Russo et al.,[20] in contrast, advocated a modified Fontan procedure as a viable alternative for this difficult group of patients. We[21] have proposed seeking to achieve biventricular repair, using the technique of translocation of the interventricular communication. We employed the technique in a 17 month-old infant with complete origin of the aorta from the right ventricle and obstruction of the subpulmonary outflow tract. Following closure of the ventricular component of the atrioventricular septal defect, we created a new ventricular septal defect in the subaortic area (Fig. 4). We then tunnelled the new defect to the aorta using a separate patch (Fig. 5). Because of the obstruction in the subpulmonary outflow tract, we used a valved homograft to re-establish continuity from the right ventricle to the distal pulmonary arteries. The patient also had congenital division of the left atrium, which was repaired simultaneously. The translocation of the interventricular communication permitted adequate growth of the outflow from the left ventricle, as evidenced by long-term angiographic and haemodynamic data showing no obstruction nine years after initial repair.

## Repair of common atrioventricular junction with discordant ventriculo-arterial connections

It is rare to find transposition associated with a common atrioventricular junction. For example, in a review of 132 patients with transposition, Boesen[22] found only 3 with a common atrioventricular junction. As already discussed, Bharati and associates[11] found 17 hearts with discordant ventriculo-arterial connections among 507 specimens of atrioventricular septal defect with common atrioventricular junction. Obstruction of the left ventricular outflow tract, either stenosis or atresia, was present in all these hearts. On the other hand, Shaher[23] collected 178 specimens with discordant ventriculo-arterial connections, but found no cases having a common atrioventricular junction.

In the rare cases having common atrioventricular junction and transposition without obstruction of the left ventricular outflow tract, the malformation can be corrected by septation of the atrioventricular junction combined with the arterial switch operation.[24–27] Obstruction of the left ventricular outflow tract, however, is present in the majority of cases, and significantly complicates the anatomic repair. If the obstruction is subvalvar and dynamic, a concomitant arterial switch with septation of the common atrioventricular junction can still be entertained. Should the subpulmonary obstruction be fixed, particularly if associated with valvar pulmonary stenosis, then this precludes the arterial switch operation. It may still be possible to achieve physiological biventricular repair, septating the common atrioventricular junction, and combining this with a Mustard or Senning operation and placing a conduit from the left ventricle to the pulmonary arteries, as reported by Alfieri and Plokker.[28] Prior to this, only a Fontan operation had been reported for this combination of malformations.[29]

To achieve anatomical biventricular repair in this setting, we propose translocation of the ventricular septal defect (Fig. 4). As in the setting of common atrioventricular junction associated with double outlet right ventricle and a non-committed interventricular communication, the aorta takes its origin exclusively from the right ventricle in patients with discordant ventriculo-arterial connections. Translocation of the ventricular septal defect, therefore, will permit anatomical biventricular repair. Following septation of the common atrioventricular junction, a new subaortic ventricular septal defect can be created, and the left ventricle can be re-routed to the aorta through an intraventricular tunnel (Figs. 4 and 5). The right

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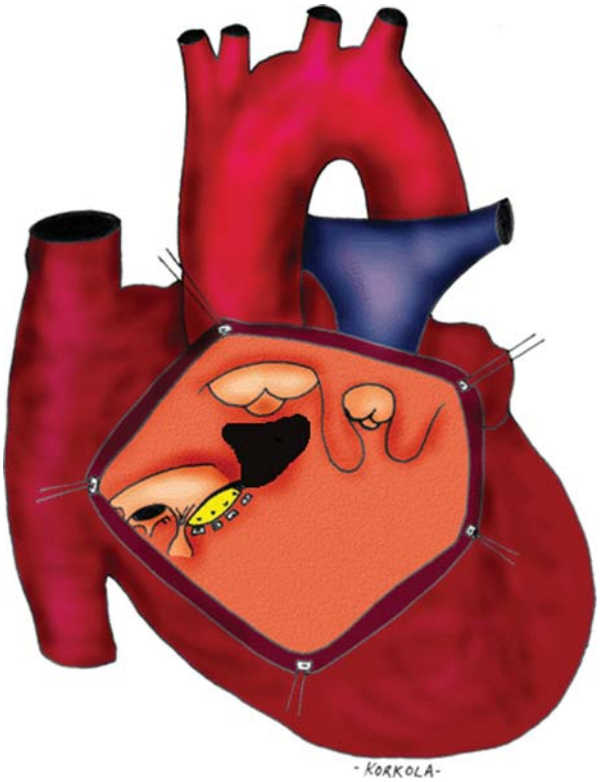


**Figure 4.**
*The cartoon shows how, after closure of the ventricular component of the atrioventricular septal defect in hearts such as illustrated in Figure 3, a new interventricular communication can be created in subaortic position.*

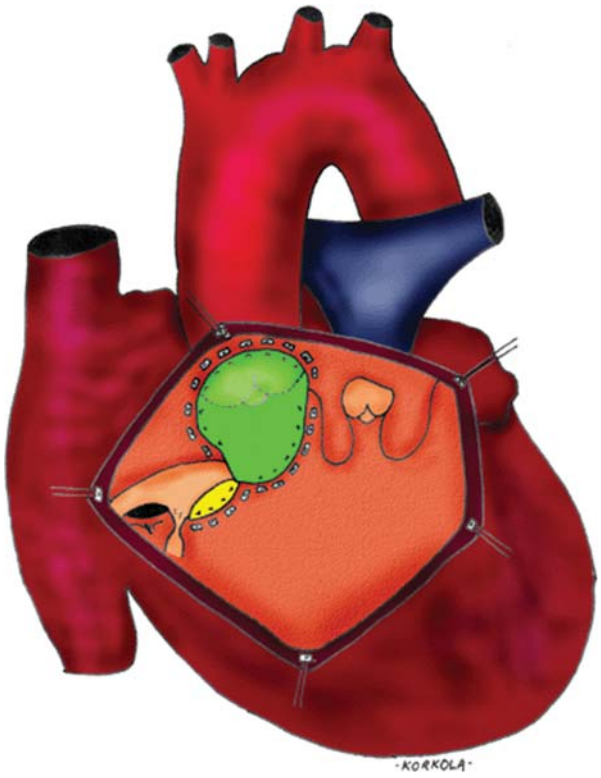


**Figure 5.**
*Having created the new interventricular communication, as shown in Figure 4, it can be tunnelled into the aorta, thus achieving biventricular anatomic repair. It is also necessary to relieve the subpulmonary obstruction (see text for discussion).*

ventricle can then be connected to the distal pulmonary arteries with a valved conduit.

## Repair of common atrioventricular junction and common arterial trunk

The association of common atrioventricular and ventriculo-arterial junctions is extremely rare. Successful surgical correction of this combination, to the best of our knowledge, was first reported by Sousa-Uva and associates in 1994.[30] The authors also reviewed the literature at that time, and found only 11 reported cases, all at post-mortem. In their patient, the superior bridging leaflet extended well into the right ventricle, type C as described by Rastelli, so that the ventricular component of the atrioventricular septal defect extended towards the truncal valve. The pulmonary arteries arose separately from the common trunk, and the papillary muscles supporting the left atrioventricular valve were fused. They split the papillary muscles, but left open the zone of apposition between the left ventricular components of the bridging leaflets. The ventricular component of the septal defect was closed via a low infundibulotomy with a Dacron patch. The common arterial trunk was transected, the pulmonary arteries detached, and a Lecompte manoeuvre performed. Continuity from the right ventricle to the pulmonary arteries was established with a valved aortic homograft. The patient was still alive three months after the operation. Subsequently, Conte et al.[31] reported the successful correction in an infant with similar anatomy, but also with totally anomalous pulmonary venous connection to the coronary sinus. Biventricular repair was accomplished in one stage at the age of 11 months, using 3 patches, 2 to repair the ventricular septal defect and reroute the left ventricle to the neoarta, and one to septate the common atrium. The common arterial trunk was transected, and the confluence of the pulmonary arteries was connected to the right ventricle with a valved homograft and a Dacron hood. The patient did well, and was noted to be in good condition 6 months post-operatively.

## Conclusions

The combination of abnormalities of the ventricular outflow tracts with a common atrioventricular junction can be found in a heterogeneous group of malformations, and presents many challenges for surgical repair. These include the construction of two competent atrioventricular valves, closure of all interatrial and interventricular defects, and construction of unobstructed

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pathways from the left ventricle to the aorta and the right ventricle to the pulmonary arteries. When the common atrioventricular junction is associated with double outlet right ventricle, the classification mirrors that used for double outlet right ventricle with separate right and left atrioventricular junctions. Biventricular repair is achievable when the common atrioventricular junction is balanced between two well developed ventricles. Repair of common atrioventricular junction in the setting of tetralogy of Fallot can be achieved either by a rightward division of the superior bridging leaflet and insertion of a single patch, or by using a comma-shaped ventricular patch to avoid subaortic stenosis in association with a separate atrial patch. This technique can also be used to repair patients with double outlet right ventricle when the ventricular component of the septal defect extends towards the aorta. Translocation of the ventricular septal defect permits anatomic biventricular repair when the defect is non-committed, or when discordant ventriculo-arterial connections are accompanied by obstruction of the left ventricular outflow tract.

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