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Realignment of the ventricular septum using partial direct closure of the ventricular septal defect in Tetralogy of Fallot☆

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Abstract

Objective: The aim is to describe our technique of partial direct closure of the ventricular septal defect (VSD) in Tetralogy of Fallot (TOF), and assess its influence on the realignment and remodeling of the left ventricular outflow tract. **Methods:** Between 2004 and 2010, 32 non-consecutive patients with TOF underwent a direct or partial direct closure of VSD. Median age and weight were 5.2 months and 6.7 kg, respectively. An approach through the right atrium was used in 30 patients and through the infundibulum in two patients. The conal septum was mobilized by transecting the hypertrophic trabeculae to facilitate the approximation of the VSD. The membranous part of the VSD was closed (in the later part of the series) with a small xenopericardial patch to avoid tension on the suture line traversing the area of risk to the bundle of His. Follow-up was complete, with a median duration of 46.9 (range 12—75.3) months. **Results:** The VSD could be closed successfully in all patients. A residual VSD was partly responsible for one early postoperative re-operation. There were no early or late deaths. At follow-up, all patients were in sinus rhythm. Three patients showed a small residual VSD. Thirty patients had none, one showed trivial, and one had mild aortic regurgitation. The left ventricular outflow showed a good realignment of the ventricular septum in all the patients. **Conclusions:** Partial direct closure of the VSD corrects the primary defect in TOF, that is, the malalignment of the septum. It results in a straight, wide open left ventricular outflow tract and brings better support to the aortic root.

Keywords: Tetralogy of Fallot; Malaligned VSD; Partial Direct VSD closure; Left ventricular outflow tract; Right ventricular outflow tract

1. Introduction

Tetralogy of Fallot (TOF) is the consequence of an antero-cephalad displacement of the conal septum. This results in a malaligned perimembraneous ventricular septal defect (VSD), overriding aorta, right ventricular outflow tract (RVOT) obstruction and consequent right ventricular hypertrophy. Conventional repair uses patch closure of the ventricular septal defect and enlargement of the RVOT. Because of the overriding aorta, the left ventricular outflow tract often shows a bayonet morphology after the insertion of a patch (Fig. 1). Late occurrence of aortic valve regurgitation has many reasons [1—4], among which distortion of the right coronary cusp by the patch and lack of support to the overriding aortic annulus may play an important role. This study describes our technique of partial direct closure of the VSD in TOF and speculates upon the potential long-term advantages.

2. Materials and methods

The study includes 32 (nine female) non-consecutive patients operated upon at University Children's Hospital Zurich with a direct closure of VSD in TOF between 2004 and 2010. This series included all the consecutive TOF total corrections performed by the same surgeon. Median age at correction was 5.2 (range 1.56—20.06) months. Median weight was 6.73 (range 3.38—9.3) kg. Two patients had a prior neonatal palliation with a modified Blalock-Taussig (BT) shunt.

The VSD was accessed through a conventional right atriotomy, a periannular detachment of the anterior leaflet of the tricuspid valve in 30 patients, and through an infundibulotomy in two patients. While a total direct closure of the VSD was performed (sometimes buttressed with a strip

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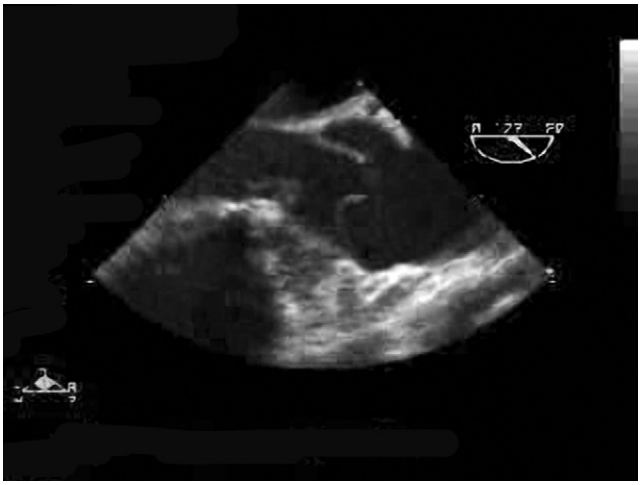

Fig. 1. Bayonet left ventricular outflow morphology with eventual sporn formation late after patch closure of a malaligned ventricular septal defect.

of pericardium) in the early part of the experience, the technique was adapted with the accumulation of experience. Today, the conal septum is first set free from the tethering trabeculae (through a small infundibulotomy) and the malaligned ventricular septum is anastomosed directly to the mobilized conal septum through a transatrial approach, using running polypropylene suture. This reduces the tension on the suture line, which is greater at the posterior part of the VSD. Instead of direct closure at the membranous part of the septum, a membranous septum is recreated using a 4–5 mm patch (Fig. 2). The detached anterior leaflet of the tricuspid valve is reapproximated using a running suture of polydioxanone [5,6].

The RVOT was enlarged with a trans-annular patch in 24 patients (12 of them with a monocusp), an infundibular patch in seven patients, and a patch on the pulmonary artery in one patient. Pulmonary valve commissurotomy was performed whenever necessary.

Clinical and echocardiography follow-up inquiries were performed at University Children's Hospital Zurich and allied peripheral hospitals. Follow-up was 100% complete with a median duration of 46.9 (12.1–75.3) months.

3. Results

The VSD could be closed directly in all patients, including those with more than a 50% overriding aorta (four patients). There was no early or late mortality.

3.1. Residual VSD

A few residual VSDs could be traced with trans-esophageal echocardiogram (TEE), especially at the beginning of the experience. On medium-term follow-up, three patients showed a small, hemodynamic ally non-relevant residual VSD. One patient was re-operated on during the same hospitalization because of a combined residual VSD and RVOT stenosis.

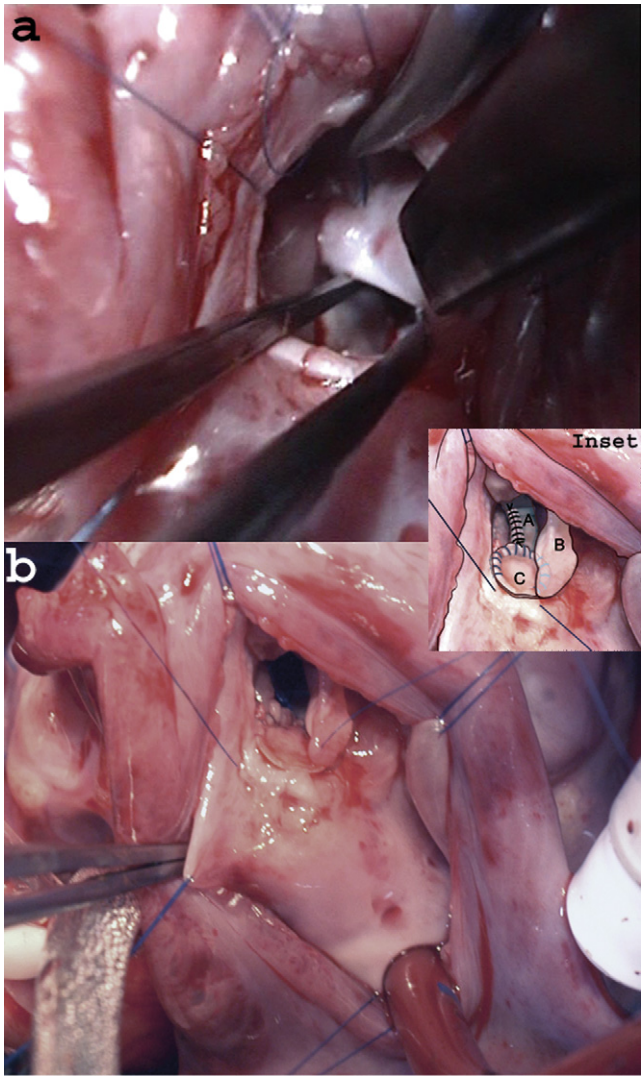

Fig. 2. Operative images showing direct adaptation of the muscular component of the malaligned VSD (a) and recreation of a membranous septum using a xenopericardial patch proximally (b). Approach is through a periannular detached anterior tricuspid valve leaflet. The inset semi schematic picture show the direct closed suture line (A), the xenopericardial patch (B) and the detached anterior leaflet of the tricuspid valve (C).

3.2. Rhythm

A few patients showed a transient third-degree atrioventricular (AV) block (mostly in the initial part of the experience), which recovered into a stable sinus rhythm within a few minutes. One patient received a DDR pacemaker due to persistent postoperative AV block III°, but has recovered since to a stable sinus rhythm.

3.3. Left ventricular outflow tract and aortic valve function

The left ventricular outflow tract has shown good realignment without evolution of a sporn or membrane formation, occasionally seen with redundant patches. Aortic valve competence improved from minimal or mild regurgitation in six patients preoperatively, to in two patients at last

follow-up. Based on a full data set of aortic annulus measurements available (15 patients), aortic annulus *Z* score changed from a predischarge median of 1.34 (range 0.7—3.71) to a median of 2.53 (range 0.61—4.06) at last follow-up. At this point of time, all but the above-mentioned two patients are free of any aortic regurgitation.

3.4. Pulmonary and tricuspid valve function

At last follow-up, 17 patients have trivial to mild pulmonary regurgitation, 13 have moderate, and two have severe pulmonary regurgitation. None of the patients has significant residual obstruction across the RVOT. While 20 patients had some form of tricuspid regurgitation preoperatively, seven had trivial, and five mild tricuspid regurgitation at last follow-up.

3.5. Re-operation and re-intervention

Three patients were re-operated during the same hospitalization. Two re-operations were already mentioned (residual VSD closure and pacemaker implantation). The other re-operation consisted of relieving a right pulmonary artery stenosis (without cardiopulmonary bypass). Two other patients underwent a late re-operation during follow-up (22.6 and 32.7 months) for insertion of a valved conduit for increasing pulmonary regurgitation. One further patient required a left pulmonary artery (LPA) stenting 8 months postoperatively.

4. Discussion

TOF is a complex congenital malformation with excellent early and late outcome after intracardiac repair [2,7,8]. With increasing numbers of patients with corrected TOF surviving into adulthood [9—11], attention is being diverted to residual lesions, hitherto considered benign, which can influence the longevity and quality of life of these patients, the aim being to achieve a survival and quality of life comparable to normal population. The integrity and function of the outlet septum (the VSD in the pre-repair TOF), the right and left ventricular outflow tracts including the valves, the tricuspid valve, and the sequential rhythm are critical to the long-term outcome. The present dispensation of repair of TOF fares well on most counts [11]. While new knowledge about the importance of pulmonary valve competence in corrected TOF has set new guidelines to ensure normal cardiac function over the long term [12,13], knowledge about the less common left ventricular outflow problems is just emerging [1—4,14]. Our technique of partial direct closure of VSD to realign the ventricular outlet septum and to re-establish the muscular scaffold to the aortic annulus attempts to recreate the natural morphology. With this technique, the outlet septum retains its contractile potential and function, in contrast to the akinesia of a patch.

While we started with a direct suture closure of the complete VSD, with experience, we learnt and introduced a few modifications.

We understood that transection of anomalous muscle bundles in the RVOT (especially those around the aortic annulus) allowed some mobilization of the conal septum. This step seems critical to our technique and has to be performed first. Second, we started using a small patch at the subtricuspid component of the VSD, where the membranous septum is supposed to be. These steps ensure a tension-free anastomosis and also avoid any plication of the tricuspid annulus. They also contribute to the protection of the His bundle. With this modification, we have no longer seen a transient AV block, which occurred sometimes in our early experience.

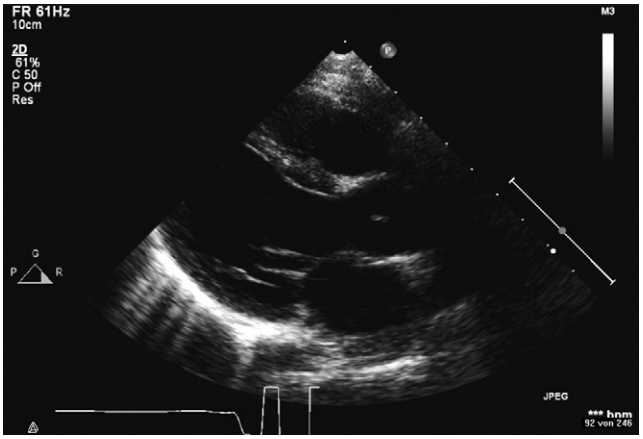

Fig. 3. A trans-esophageal echocardiography image showing the realigned ventricular septum and a harmonious left ventricular outflow 4 years after partial direct VSD closure in TOF.

As a principle, we perform a palliative shunt only when a neonatal TOF presents in blue spells; we perform a more complete correction as of 6 weeks of life. We tend to perform an elective total correction between 3 and 6 months of age. This time frame means that there is less of hypertrophy (lesser muscle resection is required), and the myocardium is more elastic. These contribute to ensure minimal stretch during primary muscle-to-muscle adaptation.

Direct VSD closure results in straightening and realignment of the outlet septum resulting in a more harmonious LVOT (Fig. 3). It does not cause a subaortic angulation, sometimes seen with a redundant VSD patch in a corrected TOF (Fig. 1). Theoretically, the posterior realignment of the conal septum should also make more room for the RVOT; however, we have no objective evidence at present to support this theory.

Buttressing the aortic annulus with a muscular septum recreates in a way how nature would have it. To predict the long-term consequences of this modification on the aortic annulus, aortic valve, and the ascending aorta would be speculative at the moment. However, it could potentially harmonize aortic annular growth resulting in improved aortic valve competence. At the least, it should avoid any distortion of the aortic annulus, as can happen when a patch induces turbulences and distorts the right coronary cusp. We have seen very good aortic valve function in this series of patients over a median 47 months' follow-up, in spite of a mild rise in aortic annulus *Z* value from 1.3 (predischarge) to 2.5 (follow-up). This contrasts with the series of Francois et al. [2], who have a higher predischarge mean *Z* value of 3.32 ± 1.66, decreasing to 0.95 ± 0.7 at 84 months of follow-up. In

addition to the retrospective quality of our echocardiography data from a select 15 patients (those which were available), the younger age at correction (5.2 vs 9.7 months), the lower proportion of shunted patients (2/32 = 6.3% vs 22%), and shorter follow-up duration, make comparison difficult. We would need to follow the aortic measurements prospectively, before drawing any conclusions.

Although direct closure of isolated VSD has been performed and reported since long [15,16], to our knowledge, this is the first report describing a partial direct closure of the malaligned VSD and realignment of the ventricular septum while correcting TOF. Our Kaplan–Meier freedom from any re-operation of 85.2 ± 13.6% at 5 years, as well as a mortality of 0%, match favorably with contemporary results [2,8].

5. Conclusion

Realigning the outlet septum while closing the VSD during repair of TOF is a novel concept that is feasible and safe. It is a work in progress and, hence, has a learning curve. It recreates a harmonious left ventricular outflow tract and buttresses the aortic annulus, with a potential to minimize subaortic problems, avoid turbulences, and facilitate better remodeling. It is an attempt at anatomical correction of the embryological basis of the malformation of TOF.

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Appendix A. Conference discussion

Dr A. Bogers (*Rotterdam, Netherlands*): If I understood correctly, it was not entirely direct closure; patches were used in some patients?

Dr Prêtre: It was complete direct closure in half of the patients, and then we have recreated this membraneous septum with a tiny patch.

Dr Bogers: I understand.

Dr Prêtre: But the purpose of that is to realign the septum. We should have called this communication 'realignment of the septum'.

Dr Bogers: Maybe we should leave it to the invited discussant to make his points and go to Dr Mignosa.

Dr C. Mignosa (*Catania, Italy*): Frankly, I must admit that I am pretty happy with the surgical results of Tetralogy of Fallot, both in the early period and long-term.

Anyhow, despite the fact that we know that aortic incompetence in the long run is, in part, due to some histological changes of the aortic wall which leads to ascending aortic dilatation and aortic incompetence, I do believe that the concept of straightening the septum is very clever, very interesting and appealing.

I have some questions: it is not that clear to me how reproducible the technique is.

Another question is, do you think that this is applicable to all patients with Tetralogy of Fallot, or do you think that in your experience you can identify a cohort of patients that could achieve more benefit with this technique?

And then, of course, I think you agree that we need more time to see the long-term results of these procedures.

Dr Prêtre: Obviously we were also cautious in introducing this technique because, as you say, the conventional approach works very well.

We always assess the elasticity of the septum, and if we have the impression it comes to the conal septum without any tension, we go ahead and do it.

Dr A. Polimenakos (*Chicago, Illinois*): Just by reviewing the video, are you doing a ventriculotomy to access the VSD?

Dr Prêtre: We closed the VSD through the atrium.

Dr Polimenakos: Ventriculotomy. So my question is, I understand the anatomical advantage of doing that, but in terms of the follow-up effect of ventriculotomy and ventricular arrhythmias, are you concerned about it? Meaning, does the toll you pay for that ventriculotomy in the long run justify this particular approach?

Dr Prêtre: It is not the issue of the Fallot pathology. In our school, we do not like pressure problems after Fallot repairs. We prefer a volume problem. That is why when we have a small hypertrophic infundibulum, we enlarge it, but we do not close the VSD through the infundibulotomy.