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Outcomes and re-interventions after one-stage repair of transposition of great arteries and aortic arch obstruction☆

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Abstract

Objectives: One-stage repair of transposition of great arteries (TGA) and aortic arch obstruction (AAO) is currently advocated, but carries formidable surgical challenges. This report presents our experience and re-interventions for residual lesions over the last 10 years. **Methods:** Twenty-two patients (19.5 ± 42.4 days; range 2–206; median 10 days, 3.5 ± 0.6 kg) diagnosed with TGA (nine patients) or double outlet right ventricle (DORV) (13 patients) and AAO underwent one-stage repair. Of the nine TGA patients (two with intact ventricular septum), AAO were: two patients hypoplastic arch, one patient discrete coarctation, four patients hypoplastic arch with coarctation and two patients interrupted aortic arch. The 13 DORV patients were all of Taussig–Bing type and one showed multiple ventricular septal defects (VSDs). The degree of AAO ranged from hypoplastic arch in five patients, coarctation two patients, combined four patients and interrupted aortic arch (IAA) two patients. Arterial switch with Lecomte ± VSD repair was performed during cooling, and aortic arch repair was performed under deep hypothermic circulatory arrest (DHCA) (35 ± 14 min at 16.9 ± 0.7 °C). Our preference was to use homograft patch-plasty for arch and direct end-to-side anastomosis for coarctation repair. Aortic-cross-clamp time was 124 ± 24 min and cardiopulmonary bypass (CPB) time 215 ± 84 min. **Results:** Early survival was 19/22 (86%) up to 30 days without mortality in the second half of our series. Three patients required extracorporeal membrane oxygenation (ECMO) support and renal support was needed in three and preferred permanent pace maker (PPM) implantation in two. Length of stay was 21.9 ± 22.1 days. There was one late death and overall survival was 18/22 (82%) for the follow-up period of 4.8 years (0.2–9.8 years). Eight patients (44%) required re-intervention for re-coarctation. Four patients required right ventricular outflow tract (RVOT)/pulmonary artery re-interventions. At follow-up, there was no requirement for aortic valve replacement, residual VSD closure and no evidence of ventricular dysfunction. **Conclusions:** One-stage repair of TGA/DORV and AAO can be performed safely with a good survival rate. Three important lessons that we have learnt are as follows: (1) the subpulmonary VSD may have a perimembraneous component, (2) late re-coarctation is not infrequent and (3) late residual right-sided cardiac lesions remain an issue in complex TGA repair.
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Keywords: Transposition of the great arteries; Taussig–Bing anomaly; Aortic arch obstruction; Arterial switch

1. Introduction

Taussig–Bing anomaly (TBA) is a well-defined entity with double outlet right ventricle (DORV), subpulmonary ventricular septal defect (VSD) and malalignment of the outlet and main inter-ventricular septum. The latter might contribute to the development of aortic arch obstruction (AAO) by preferential flow from the left ventricle into the pulmonary artery. It is not surprising to find a frequent association of TBA and AAO in the form of a hypoplastic aortic arch as previously described present in about 50% [1,2] of patients. By contrast, this combination is rare in the transposition of great arteries (TGA) complex reported in about 6% of cases. The natural course of both complex malformations does carry a very poor natural history and the treatment strategy is difficult [3,4].

One-stage repair in the setting of TGA or DORV with subpulmonary VSD and AAO has been generally accepted as proprietary surgical strategy at experienced centres. However, the lengthy surgery includes the arterial switch operation (ASO), aortic arch repair with VSD closure, which can be complex, can be challenging, and it can further be compounded by complex coronary anatomy, the discrepancy of the great vessels and the possible presence of sub-aortic obstruction.

This rare and complex congenital anomaly presents an important surgical challenge and harbours the risk for re-interventions. Nevertheless, the operative mortality has decreased in the current practice [5,6], but there is still a

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significant risk of late re-interventions such as re-coarctation, late aortic regurgitation, right ventricular outflow tract (RVOT) re-obstruction, and late (and often silent) coronary distortion, especially in the context of complex coronary transfer. This publication looks primarily at early outcomes and postoperative re-interventions after one stage repair over the last 10 years at Great Ormond Street Hospital.

2. Patients and methods

2.1. Patient population

Complete one-stage repair through midline sternotomy was performed in 22 patients with TGA complexes and AAO from 1999 to 2009 at Great Ormond Street Hospital. All underwent ASO ± VSD closure and AAO repair. The local Research and Development committee approved the study design and waived the need for individual consent taking.

2.2. TGA with AAO group

This group included nine patients, and the diagnosis was established with echocardiography in all patients. A balloon atrial septostomy (BAS) was performed three times and six patients required prostaglandin infusion to improve the distal perfusion via the ductus arteriosus. The great vessel arrangement was antero—posterior in six or side-by-side in three patients. The degree of AAO ranged from discrete coarctation to hypoplastic transverse arch or complete interruption. We used the Melbourne Children's Hospital group's criteria [7] of AAO; aortic arch diameter less than the weight of the baby + 1 mm. The AAO included: 2/9 hypoplastic arch, 1/9 discrete coarctation, 4/9 hypoplastic arch with coarctation and 2/9 interrupted aortic arch (see Table 1).

Coronary patterns are described using the Leiden Classification (see Table 2). Three of the nine neonates showed usual coronary configuration (1LAD, circumflex coronary artery (Cx); 2right coronary arteries (RCA)) with a juxta-commissural left coronary artery (LCA) in one. Abnormal patterns were identified in six. Two patients had (1LAD; 2RCA, Cx), one inverted origins (1RCA; 2LAD, Cx), one (1LAD, RCA; 2Cx), one (1 Cx; 2RCA, LAD) and one patient had separate origin of two coronaries from the same sinus (2RCA; 2LAD, Cx) along with both juxta-commissural position and intramural course of the LCA.

Two patients had an intact ventricular septum. The interventricular communication was peri-membranous in six patients and one patient had multiple VSDs. The pulmonary artery was described as being more than 50% larger than the aorta in six patients and, in three patients, the sized discrepancy was reported as non-significant.

At operation, the median age of the two girls and seven boys was 10 days (range 3— 17 days) and weight was 3.5 ± 0.3 kg.

Table 1
Preoperative data.

	DORV, AAO	TGA, AAO	Overall
Overall	13	9	22
VSD	13 (100%)	7 (77%)	20
Hypoplastic arch	5 (38%)	2 (22%)	7
Coarctation	2 (15%)	1 (11%)	3
Hypoplastic arch/coarctation	4 (30%)	4 (44%)	8
IAA	2 (15%)	2 (22%)	4
Usual coronary pattern	5 (38%)	3 (33%)	8
Unusual coronary patterns	8 (62%)	6 (66%)	14

Table 2
Coronary anatomy distribution in Leiden classification (LAD: left anterior descending artery, CX: circumflex artery, RCA: right coronary artery).

	TGA	TBA	Overall
1LAD, Cx; 2 RCA	3	5	8
1LAD; 2RCA, Cx	2	4	6
1RCA; 2LAD, Cx	1	2	3
2RCA; 2LAD, Cx	1	0	1
1LAD, RCA; 2Cx	1	1	2
2LAD, RCA, Cx	0	1	1
1Cx; 2RCA, LAD	1	0	1
Overall	9	13	22

2.3. DORV with AAO group

Of the 13 DORV, all were of Taussig—Bing anomaly as diagnosed by echocardiography. BAS was necessary in one patient and prostaglandin needed in nine patients. The great vessel arrangement was antero—posterior in four patients, oblique in seven patients or side-by-side in two. The degree of AAO ranged from hypoplastic arch in 5/13, discrete coarctation in 2/13, both hypoplastic arch and coarctation in 4/13 and interrupted aortic arch (IAA) in 2/13.

The majority (8/13) had an unusual coronary pattern: four patients with (1LAD; 2RCA, Cx), one (1LAD, RCA; 2Cx), two patients had inverted origins (1RCA; 2LAD, Cx) and one single coronary (2LAD, RCA, Cx). Usual coronary configuration (1LAD, Cx; 2RCA) was present in five out of 13. All VSDs were subpulmonary with septal malalignment. Seven patients had a significant size mismatch between the larger pulmonary artery (>50%) and aorta. However, only one patient was found to have significant preoperative right ventricular outflow tract obstruction (RVOTO) gradient.

Age at operation of the four girls and nine boys was 26 ± 54.8 days (range 2—206 days) and weight 3.6 ± 0.7 kg.

3. Methods

3.1. Surgical management

The sequence of procedures might vary according to surgeons' preference, but the principle of our ASO for TGA/VSD and TGA/DORV was similar to that used in our institution for other neonates with TGA [8]. With concomitant repair of AAO, surgery was performed with cardiopulmonary bypass using profound hypothermic circulatory arrest and pH stat strategy. Marker sutures were placed on the original pulmonary artery to facilitate coronary transfer, keeping in mind that, in TGA, the origins of coronary arteries are at a higher level than usual.

Additional ductal cannulation was required in cases of IAA. In the case of TGA and intact ventricular septum, single venous cannula was used. Bicaval venous cannulation was adopted for those with VSD, which would be usually closed

through the right atrium during cooling. Cold-blood cardioplegia and venting of the LV through right pulmonary vein was performed. The original aorta would be divided high to facilitate the Lecompte. The technique of coronary transfer, most often, involved the use of medially hinged trapdoor incisions in the neoaorta. The trapdoor incisions would not be over-generous as they could distort the sino-tubular junction. One patient with an intramural RCA and LCA arteries from sinus 2 required detachment of the posterior commissure, unroofing the stenosed orifice and a single-button transfer with a pericardial hood.

In the case of DORV, the harvesting of coronary buttons from the smallish aorta facilitated the exploration of the RVOT. Any prominent parietal band (such as in tetralogy) would be routinely divided through the aortic valve. No right ventricular (RV) incision or infundibular patch was required. The unusual coronary configurations were managed using the medially hinged trapdoor technique except for the one with the double ostium from sinus 2. The VSD was closed (left ventricle (LV) to neoaorta) with either a bovine pericardial or Gore-Tex patch. The approach was through the right atrium (RA) in eight patients and, in four patients, the VSD closure was through a combination of right atrial and the original pulmonary valve. With the latter approach, the upper edge of the VSD (subpulmonary conus) is masked by the overriding pulmonary artery annulus. This could be unfolded using a gentle push of the outer ventricular wall and the upper margin of the VSD was exposed for suturing. The VSD patch was secured using a 7/0 Prolene suture with a 6.5 mm semicircle needle, without interfering with the anterior neoaortic annulus and cusps. In addition, the VSD patch was trimmed without excessive material, which could impinge upon the neoaortic valve.

For concurrent coarctation/hypoplastic arch repair, the approach varied across the passage of time. In principle, the ductal tissue was excised and the well-mobilised proximal descending aorta was attached to the junction of the equally well-mobilised distal ascending aorta and proximal transverse arch in an end-to-side fashion (see Fig. 1), or in the case of discrete coarctation, a direct end-to-end connection would be adequate. The use of Lecompte manoeuvre also relives the tension on the repaired arch. For the interrupted aortic arch repair, ductal tissue was excised and a posterior hemi-anastomosis was performed between the distal arch and the posterior wall of the proximal descending aorta. The arch reconstruction was then augmented with a homograft patch (if available) for treating the neoaortic/original aortic size mismatch and aortic arch hypoplasia (see Fig. 2).

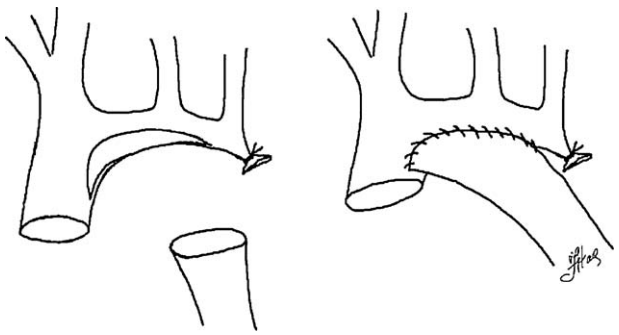

Fig. 1. Surgical technique for hypoplastic arch repair: after excision of ductal tissue the proximal descending aorta is attached to the junction of the distal ascending aorta and proximal transverse arch in an end-to-side fashion.

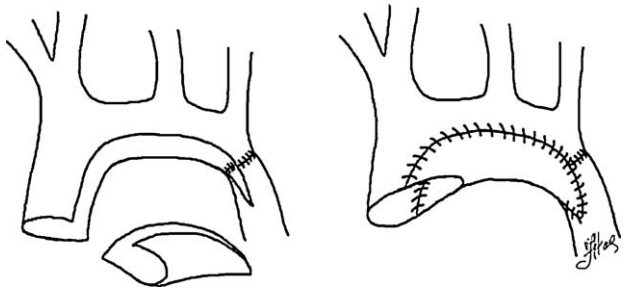

Fig. 2. Surgical technique for the interrupted aortic arch repair: After complete removal of ductal tissue, a posterior hemi-anastomosis is performed between the distal arch and the posterior wall of the proximal descending aorta. Followed by augmentation of the arch reconstruction with a homograft patch to compensate for size incongruence. Accommodating for the new aortic arch configuration after the Lecompte maneuver and to avoid kinking of the mid-aortic patch, it is of utmost importance to tailor the patch in a short fashion at its concave portion.

In 12 DORV patients, the aortic arch was reconstructed with use of a pulmonary homograft patch, and in one, the repair was performed with a Shelhigh bovine curved pericardial patch. In the TGA group, four different techniques were used for arch repair. First, the pulmonary homograft patch augmentation was used in five patients and in one with a bovine pericardial patch. Extended end-to-end and end-to-side repair in one each and reversed left subclavian flap combined with homograft patch enlargement in one patient (Table 3).

The Lecompte manoeuvre was used in all cases. Despite the great vessel discrepancy, neoaortic anastomosis was always performed without any added material to preserve the orientation of the transferred coronary buttons and sino-tubular junction. One should be cautious in the case of homograft patch enlargement of the aortic arch; the proximal inner curvature would need to be shortened to prevent the creation of an infolding at the now posteriorly sited neoaortic anastomosis.

Ultrafiltration on bypass to maintain haematocrit more than 30% and modified ultrafiltration after bypass was used. A left atrial pressure monitoring line and peritoneal dialysis catheter were inserted intra-operatively. The chest might be left open at the end of the procedure at the discretion of the surgeons.

Table 3
Intra- and peroperative data with special emphasis on aortic arch obstruction repair.

	DORV, AAO	TGA, AAO	Overall
Arch repair using a Shelhigh bovine aortic curved patch	1 (8%)	0	1
Arch repair using a bovine pericardial patch	0	1 (11%)	1
Arch repair using a pulmonary homograft patch	12 (92%)	5 (55%)	17
Reversed LSC flap + pulmonary homograft patch	0	1 (11%)	1
End-to-side anastomosis	0	1 (11%)	1
End to end anastomosis	0	1 (11%)	1
Lecompte	12 (92%)	9 (100%)	21
Delayed stenal closure	9 (69%)	6 (66%)	15
ECMO	2 (15%)	1 (11%)	3

The mean period of deep hypothermic circulatory arrest time was 35 ± 14 min, aortic-cross-clamp time was 124 ± 24 min and cardiopulmonary bypass time was 215 ± 84 min.

3.2. Postoperative care

Postoperative care included inotropic support and liberal use of vasodilators with the aim of maximally reducing the afterload. Our current preferred regime is a combination of adrenaline (0.05—0.1 $\mu g\ kg^{-1}\ min^{-1}$) and milrinone (0.25—0.75 $\mu g\ kg^{-1}\ min^{-1}$). Excessive volume loading is avoided. Lower than usual systemic blood pressure is accepted, provided the systemic perfusion and metabolic status are maintained. In case of low cardiac output not responding to the above strategy, core cooling, paralysis and peritoneal dialysis are used. Mechanical support with extracorporeal membrane oxygenation (ECMO) is used if conventional measures fail, as was required in three patients.

3.3. Follow-up

Mean follow-up time was 4.8 years (0.2—9.8 years). Primary data were collected, retrospectively, from clinical notes and the digitalised patient database. All follow-up information complemented with direct contact with the patients' care taker, as well as, if required, the patients' paediatrician or external cardiologist. All follow-up data were jointly reviewed by a cardiologist and a cardiac surgeon.

All re-interventions, whether percutaneous or surgical, for either re-coarctation or RVOT were identified. The three hospital deaths were excluded from the re-intervention follow-up.

Data are described as frequencies, means with standard deviation and means with range where adequate. Time-related survival and freedom from re-intervention and operation were calculated using the Kaplan—Meier method with GraphPad Prism version 3.02.

4. Results

4.1. Early mortality

Out of 22 patients, there were three early deaths (13.6%), all of them from the TBA patient group.

Patient 6: TBA, (1LAD; 2RCA, Cx), oblique relationship of the great vessels with significant discrepancy 2:1. Difficult LAD due to short mainstem and early trifurcation. After revision of the LAD anastomosis, failure to wean from bypass required ECMO support. This was discontinued early because of no myocardial recovery.

Patient 7: TBA with inverted coronaries (1 RCA: 2LAD, Cx), side-by-side relationship of great vessel with mild great vessel discrepancy. Patient underwent uneventful surgery but went into renal failure and died of sepsis on postoperative day 14.

Patient 11: TBA with inverted coronaries (1RCA; 2LAD, Cx), side-by-side great vessels and hypoplastic aortic arch. Failure to wean from bypass due to poor bi-ventricular function, the child was commenced on ECMO. Postoperative angiogram showed a satisfactory coronary transfer, but there was a residual VSD, tortuous aortic arch repair with severe stenosis and poor flow into the LPA. During re-operation, the Lecompte was taken down and the hypoplastic arch reconstructed with a bovine pericardial patch. The MPA was placed posterior to the neoaorta. Two days after re-operation, the child was weaned off ECMO, but soon after, experienced a sudden cardiac arrest requiring mechanical resuscitation. Despite prompt reinstitution of ECMO support, the patient was diagnosed with severe brain injury and therapy was withdrawn 17 days after initial surgery.

4.2. Late death

The single late death was the only mortality in the TGA group. Patient 10 (1Cx: 2RCA, LAD), with antero—posterior relationship of the great vessels had an uneventful surgery. There was a sudden haemodynamic deterioration during transfer to intensive care unit (ICU). Prompt initiation of ECMO was undertaken that was subsequently successfully weaned on postoperative day 6. Angiogram on day 15 showed LPA stenosis, which was not amendable to ballooning/stenting. There was no coronary distortion. The patient developed mediastinal wound infection and renal insufficiency requiring dialysis. The patient subsequently died 112 days postoperatively due to sepsis.

The four deaths were all within the first half of the described experience. There was no early or late mortality for the second half of the study.

4.3. Early morbidity

Other operations that became necessary were for permanent pacemaker implantation in two patients in the DORV group (9%), for hemidiaphragm plication in two patients (9%) and for delayed sternal closure in 15 patients (68%). Ventilatory support was for an average of 10.8 ± 22 days. Hospital stay was 21.9 ± 22.1 days.

4.4. Late re-interventions

Re-interventions and re-operations became necessary for recurring re-coarctation or right-sided obstruction in nine of 18 surviving patients (five of TGA and four of the DORV group) within the first year post-surgery. Only one patient underwent re-operation for isolated pulmonary artery stenosis. The remaining eight patients required balloon angioplasty for moderate recurrent AAO in the first instance. Three out of the eight needed a second arch reconstruction for recurrent AAO. In one, a subclavian flap repair was undertaken and in two homograft or Gore-Tex patch-plasty was performed.

After undergoing aortic arch re-operation, three patients required a third time operation for pulmonary artery stenosis (two of TGA and one of DORV group) and one TGA patient for RVOT obstruction. The latter was related to a smallish neopulmonary valve with a right coronary artery crossing the ventriculo—arterterial junction. Repair was performed with a Gore-Tex conduit from the RV infundibulum to the pulmonary trunk as a double-barrel technique. Of the pulmonary artery

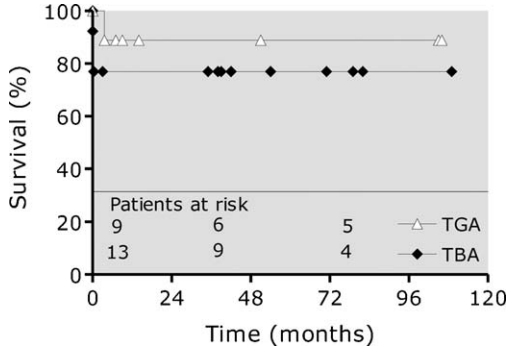

Fig. 3. Actuarial survival.

stenosis, one was relieved with a homograft patch-plasty of the pulmonary trunk and two with Gore-Tex patch-plasty of the pulmonary bifurcation associated with the main pulmonary trunk.

4.5. Follow-up

Of the 18 survivors, 16 had complete echocardiographic/clinical follow-up data, and the functional status was the New York Heart Association (NYHA) class 1. Eleven aortic valves and six pulmonary valves showed trivial to mild regurgitation. Biventricular function was well preserved and no valvular replacement was required. Maximum aortic arch flow velocity was measured at 2.1 ± 0.3 m s^{-1} and the flow velocity over the RVOT was 1.8 ± 0.3 m s^{-1}. Branch pulmonary artery (PA) flow velocities were 2.5 ± 0.8 m s^{-1} for right pulmonary artery (RPA) and 2.3 ± 0.7 m s^{-1} for the left pulmonary artery (LPA).

Overall survival was 82% for the follow-up period of 4.8 years (0.2—9.8 years). Actuarial survival for TGA and TBA, respectively, are 89% and 77% at 10 years (see Fig. 3). There is no linear learning curve from the beginning of this series, but the four deaths were all within the first half of the described experience. There was no early or late mortality for the second half of the study. The actuarial freedom from re-intervention among survivors over 10 years was 50% for the TGA group and 57% for the TBA group, respectively (see Fig. 4).

5. Discussion

One-stage repair of combined aortic arch obstruction and transposition complexes presents a true surgical challenge.

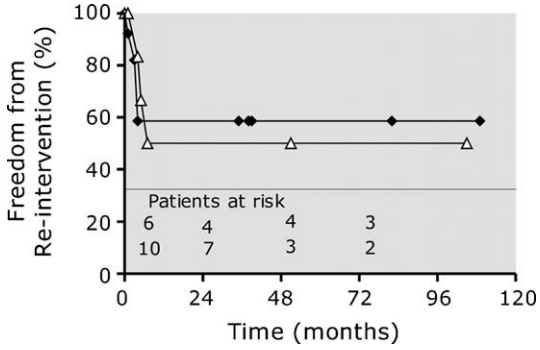

Fig. 4. Actuarial freedom from re-intervention.

Previous publications have shown the superiority of the one-stage repair compared to a two-stage strategy [5,6]. The two-stage strategy in case of a lateral approach to the arch anomalies and pulmonary artery banding carries the risk to induce, for example, neoaortic valve regurgitation [9] and branch pulmonary artery distortion and stenosis or hypertrophy of the future systemic ventricle. Without banding, the need for the second-stage operation might be earlier than anticipated because of worsening congestive cardiac failure.

The advantages of the one-stage complete repair might result from the early reestablishment of physiologic conditions and the decreased response to the PA band and by avoiding early re-operation. The arch hypoplasia, interruption or coarctation is efficiently dealt with at the time of the ASO while working through a median sternotomy. It is generally possible to perform a direct anastomosis for interruption or coarctation with mild tension on the anastomosis [10,11]. On the other hand, the aortic arch hypoplasia requires an additional longitudinal patch-plasty. This has the advantage of enlarging the size of the ascending aorta, thus facilitating a tension-free neoaortic anastomosis; it achieves an aortic arch of adequate size.

5.1. Early outcomes

To successfully undertake the complex procedures together (arterial switch, VSD repair and aortic arch reconstruction, and in the case of DORV, with great vessel discrepancy, RVOT resection and frequent unusual coronary configurations) would require the perfect transfer of the coronary arteries. In many instances, this remains a major challenge in the context of unusual coronary artery anatomy with a dilated PA in a side-by-side arrangement. In the article by Pocar et al. [12], complex coronary anatomy remains an important risk factor for this very difficult group of patients. However, in our series, transfer of unusual coronary patterns did not appear to be the direct cause of deaths. Of all four patients who died, three had abnormal coronary artery pattern — two with total inverted coronary artery anatomy (1RCA; 2LAD, Cx) and one was partially inverted (1Cx; 2LAD, RCA). Although their coronary transfer appeared to be satisfactory, two patients died of renal failure and sepsis and one had the treatment withdrawn because of brain injury. However, in the one with the less complex coronary anatomy (1LAD; 2RCA, Cx) the transfer of LAD, which had a short mainstem and early trifurcation, needed revision at the time of surgery without success and this was the direct cause of death.

5.2. Heart block

Early in our experience, the seemingly muscular margin of the subpulmonary VSD inspected through the outlet valve was probably the reason for the heart block in two of our patients post repair. We would always undertake an evaluation of the VSD via the right atrial approach in the first instance. This showed the presence of any perimembraneous component and would decrease the risk of heart block.

5.3. Late re-intervention and re-operations

5.3.1. Recurrent aortic coarctation

In contrast to some previous studies [12,13], there is an overall important need for re-intervention in half of the patients in our series. A similarly high rate for overall re-interventions of close to 43% was reported by Planché's group [9] with 19/67 patients (28%) requiring re-intervention for recurrent coarctation and 10/67 patients (15%) for neoaortic regurgitation.

In our study, a non-significant difference was found between the TGA and TBA anomalies. In the TBA group, all patients underwent pulmonary homograft patch enlargement of the aortic arch. In the TGA group, three patients underwent: one an extended end-to-end, one an end-to-side and one a reversed left subclavian flap repair complemented with a homograft patch. Only the last patient of the three required isolated re-intervention for recurrent arch stenosis. All others were patch enlarged in the same fashion as for the TBA group. The technical difference does not explain the higher need for re-intervention in the TGA patient population. Our unit preference would be to use a homograft patch to achieve an even-sized reconstruction of the aorta. However, bovine pericardial curved patches were used in two of the patients when homograft were not available. At re-operation, the thickened surface of the curved bovine pericardial patch looked 'thrombogenic' and its unpredictable behaviour might have contributed to the early fibrosis and arch re-obstruction.

Of the nine patients needing re-interventions, eight patients underwent percutaneous therapy at first. Following balloon dilatation for recurrent coarctation, surgical repair was required in three patients subsequently, including two patients repaired with a curved bovine patch in the first instance.

The heterogeneity of AAO in our series mandates individual management. Clearly, for some patients, the decision of when and how to re-intervene is difficult. Careful quantitative echocardiographic assessment can assist decision making, but for the vast majority, the decision was made during diagnostic catheterisation. At our unit, a considerably low threshold for balloon dilatation of recurrent or residual aortic arch obstructive lesions might be one explanation for the high re-intervention rate.

5.3.2. Late RVOTO

A higher prevalence of initial and subsequent RVOTO was described in a similar series [12]. We adopted a routine division of prominent subvalvular trabeculations through the original aortic valve. This would open up the RVOT adequately, in most cases. None of our patients' RVOT was patch enlarged. It is also important to mention that patching of the RVOT is often precluded by the distribution of the coronary arteries. Only one patient in our series underwent re-operation due to RVOTO related to unusual right coronary artery anatomy and the double-barrel technique has been mentioned earlier.

5.3.3. Neoaortic valve regurgitation

So far, in our limited follow-up, 11 patients showed trivial or mild aortic valve regurgitation. No re-operation was necessary for neoaortic valve regurgitation. This is in contrast to the report from Mohammadi et al. [9]. Surgical factors were thought to be contributing to the high incidence of aortic valve regurgitation and replacement. Our approach to do subpulmonary VSD through the original pulmonary valve was described in the surgical section. A gentle push of the outer right ventricular wall unfolded the upper margin of the VSD for placement of continuous suture. The VSD patch was carefully trimmed so that no excessive material impinged upon the neoaortic valve cusps. In addition, to minimise any major distortion of the integrity of the newly reconstructed sino-tubular junction, we avoided additional material in the reconstruction of the neoaortic/aortic anastomosis. The transection of the original PA is important to maintain a sino-tubular junction. Too proximal the transection below the expected level of future sino-tubular junction of the neoaorta may interfere with the valve integrity. Another aspect in the configuration of the sino-tubular junction would be to avoid oversizing of the medial trapdoor incisions.

5.3.4. Outcome analyses

It is also of interest that some of the previous studies with the same surgical therapy of ASO, VSD closure and AAO relive faced different postoperative outcomes with overall good survival. Vouhé's group [12] described a 76% actuarial survival rate in 38 patients, mentioning cardiac re-operations in five patients, two for RVOTO repair, two for left main coronary stenosis and one for a combination thereof. However, only three patients with recurrent coarctation were successfully treated with balloon angioplasty. By contrast, Planché's group published their follow-up results with emphasis on surgical factors and left-sided lesions. In this study, 67 patients underwent one-stage repair with either end-to-end (35 patients) or patch enlargement of (32 patients) for AAO relive [9]. Overall, actuarial survival in this series was 94% in the 32 patients and 75% in the 35 patients over 10 years. Re-interventions became necessary for re-coarctation in 15 patients (22%). The second most frequent cardiac reason for re-intervention was neoaortic regurgitation, a postoperative complication less described by the other discussed studies. In their single stage approached group, 55% of patients had neoaortic regurgitation.

Those results illustrate that even in the most specialised centres, postoperative complications can differ for unclear reasons that might include anatomic features of the patient samples, differences in surgical techniques, variability in the threshold and use of interventional techniques.

The one-stage-repair of transposition complex and aortic arch obstruction repeatedly showed good survival results for these highly complex congenital anomalies in high-volume hospitals. Nevertheless, this sort of surgical therapy is highly challenging and faces many intra- and postoperative hurdles. In those neonates with significant co-morbid conditions [14] such as necrotising enterocolitis or brain injury, a palliative two-stage approach should be considered. Nevertheless, in those patients with IAA and severe arch hypoplasia, the surgical option would remain very limited.

5.4. Limitations

This study represents a relatively small patient sample (22) and does limit statistical analysis. Of course, the presented

series consists of an inhomogeneous patient selection through a surgical technique rather than a diagnostic code and results can only, if at all, be compared from a surgical point of view. A selection bias might be introduced because of differences in management strategies.

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Appendix A. Conference discussion

Dr R. Jonas (*Washington D.C., USA*): I notice from your manuscript, which you kindly supplied to me, that you used no subannular patches in the neopulmonary outflow or transannular patches, which I think differs from the experience of other reported series and certainly our own experience.

How would you decide whether you needed a subannular patch, an infundibular patch, in this setting?

Dr Huber: In this setting, it is difficult to know if, as you have mentioned, subpulmonary intervention is needed.

From our early isolated TBA experience, we learned that patients required division of subvalvular trabeculations. And we became very liberal by approaching the supravalvular apparatus by dividing, resecting if necessary, the trabeculations in order to open up the outflow tract.

This might be one of the rationales of why we had so few problems. There was only one patient, as I mentioned in the presentation, requiring the baffle technique from the infundibulum onto the pulmonary artery. Because of crossing coronary artery, we couldn't create a patch reconstruction. All the others were fine, just with subpulmonary or subvalvular trabeculation division.

Dr Jonas: Another question also relates to the pulmonary outflow reconstruction. You pointed out that there is a very high incidence of unusual coronary distribution in this setting, and frequently that involves a right coronary artery that runs right over the pulmonary annulus so that if you did need an infundibular or, particularly, a transannular patch, then you are unable to do that, and you're committed to an RV-PA homograft conduit.

I wonder if you've considered in that setting, as we had to just a couple of weeks ago by chance, doing a Yasui type procedure; in other words, you would baffle the left ventricle to the pulmonary trunk, divide the pulmonary artery, and then perform a Norwood type reconstruction of the ascending-to-descending aorta and then complete the reconstruction with an RV-PA homograft.

I recommend this approach to you. I wonder if you've come across that situation?

Dr Huber: No, we have not come across it. In the 10 years we've used it, there were no strategies similar to that described.

Dr Jonas: That leads into my third point, which is that you had a very high incidence of late re-coarctation. One lesson that I've learned the hard way over the last few years is that in this particular situation where you perform an arterial switch and move the ascending aorta posteriorly to connect across an interrupted aortic arch, you now have an extremely acutely angled ascending-to-descending connection.

And in that setting, I have found it very difficult to place a single curved patch that runs from the ascending, along its full length, across the anastomosis, and then down the descending. It's so acutely angled that it's very likely to kink at the peak. And by doing the Norwood type reconstruction, specifically the Yasui operation, I've found that I've been able to avoid that problem.

Another way of getting around the angulation problem is to use two separate patches, one going up the ascending, one going down the descending, and connecting the two patches together.

So I am wondering if you could comment as to why you feel you had such a high incidence of arch problems. I believe it was something like a 45% incidence of arch recurrent obstruction.

Dr Huber: Yes, you're absolutely right. The reason for the higher need for re-intervention on the aortic arch might on one side be caused by a low threshold of the interventional cardiologist to go for a perfect result. Gradients of more than 20 mm Hg were ballooned if possible. That might be one explanation.

The second explanation, as you pointed out, is the use of the pulmonary homograft patch in the fashion we have used it and by the posterior movement of the aorta to become an acute angle.

So what we have done towards the last part of our experience is to cut the homograft patch in the inner curvature of the aortic arch in order to allow tension free but also a smoother curve again that has less of an acute angle.

I would also like to point out briefly that in our experience, we had eight patients undergoing re-intervention for re-coarctation, but only three really underwent surgical re-operation. And of those three, two were the ones where we used the bovine pericardial patches.

Dr J. Comas (*Madrid, Spain*): I have a comment myself. It's related to the aortic insufficiency. I have read your paper. That is one of the problems, mostly related sometimes to the discrepancy of a huge neoaorta with probably a small ascending aorta originally. I don't know. I understand that probably when you repair the coarctations, you go down to the ascending aorta?

But the other thing that you're saying is about the coronary reimplantation. You're using a trap door, and you're trying to reduce your technique.

I've been using trap door all the time, all of my life, in all the transpositions, and suddenly I am thinking a lot about this situation. And in this

situation, I changed to do the closed system. I'm doing the aorta first. I'm copying the model of John Brown, who luckily is here, and I was very happy. It was very successful, and the aortic valve was finished with non-regurgitation. It probably is a system to avoid this. What do you think?

Dr Huber: We had 11 aortic valves that showed trivial to mild regurgitation, but no need to reintervene on those valves. I think, therefore, the Great Ormond Street Hospital experience using the trap door technique has been shown to work out at the institution.

And as I showed on the earlier slide, the technique we used was really to take care not to be over-generous on the incisional side as well as to create a double fold in order not to increase the size further and keep the size mismatch between the neoaortic or pulmonary root and the ascending aorta as small as possible.

We also aimed at putting the coronaries high up and far away from the valve in order not to distort any valve geometry further.

So it's difficult to give you an answer, but the results speak for themselves, I think.

Yes, your second question was? Sorry.

Dr Comas: It's okay.

Dr T. Tlaskal (*Prague, Czech Republic*): Just one comment. We have an experience with 11 patients with these settings but we are using a little bit different approach for aortic arch reconstruction. The same method as in the interrupted aortic arch, that means direct anastomosis between the descending and the ascending aorta, was always used. So far we have not observed any re-coarctation.

Editorial comment

Outcomes and re-interventions after one-stage repair of transposition of the great arteries and aortic arch obstruction

Keywords: Transposition of the great arteries; Aortic arch obstruction; Arterial switch operation

Huber and collaborators report their 10-year experience with one-stage repair of transposition of the great arteries (TGA) associated with aortic arch obstruction (AAO) and (in most cases) ventricular septal defect (VSD) [1]. The results were outstanding in terms of survival (82% overall survival and no mortality in the second half of the experience). Late survivors had a satisfactory clinical outcome, but a high proportion of patients required re-operation/re-intervention for recurrent aortic arch obstruction and/or stenosis of the neo-pulmonary outflow tract.

In patients with TGA/VSD/AAO, the superiority of a one-stage repair over a two-stage approach has been suggested many years ago [2]. The present article provides further support for this superiority. The advantages of a one-stage strategy are multiple: early re-establishment of physiologic conditions, avoidance of the drawbacks of pulmonary banding and optimal repair of AAO. There is now a clear evidence that one-stage repair is the approach of choice in this complex malformation.

Perfect surgical repair represents the key point for successful early and late outcomes. This carries formidable surgical challenges. The present article illustrates most of these difficulties.

(1) Optimal repair of the aortic arch is mandatory.

Even if extended end-to-end or end-to-side anastomosis may sometimes be satisfactory, extended patch aortoplasty (as illustrated in the Fig. 2 of the present article) should be recommended. It provides even-sized reconstruction of the aorta, corrects the size discrepancy between the proximal great arteries (thus facilitating the arterial switch procedure) and may reduce the risk of recurrent obstruction (even if this last point was not demonstrated in the current study). The optimal patch material remains controversial. Heterologous pericardium may become fibrotic; pulmonary homograft may calcify; both materials have no growth potential and may increase the risk of recurrent obstruction. It has been shown that glutaraldehyde-treated autologous pericardium provides harmonious growth of the aortic arch without aneurysmal dilatation [3]. This is our material of choice.

(2) The arterial switch operation (ASO) is often technically challenging.

Several anatomic features may induce surgical difficulties: size discrepancy between the great arteries, variable relationship between the great vessels and frequent occurrence of unusual coronary patterns.

The necessity of anterior translocation of the pulmonary bifurcation (Lecompte maneuver) should be discussed. The Lecompte maneuver is mandatory when the ascending aorta is anterior to the main pulmonary artery (even in oblique relationship); it is unnecessary and potentially dangerous (coronary compression) when the great vessels are strictly side by side, and it should therefore be avoided. This technical point was, at least in part, responsible for one of the early deaths in the present series.

Unusual coronary patterns are common. In the present study, there was no negative impact of the coronary anatomy on the outcome. However, in the absence of routine coronary evaluation, the true incidence of coronary events cannot be determined. It is well established that coronary obstructions may be detected in patients without any clinical, electrocardiographic or echocardiographic evidence of myocardial ischemia [4]. We strongly recommend to evaluate