The anatomic repair of recurrent aortic arch obstruction in children and adolescents

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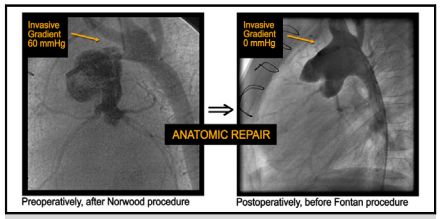
ABSTRACT

Objective: Surgery for recurrent aortic arch obstruction is highly challenging and publications are rare. The aim of this retrospective, single-center study was to evaluate mortality, complications, and reintervention rate after an anatomic repair.

Methods: Between 1999 and 2022, in total 946 operations on the aortic arch were performed at the Children's Heart Center Linz. In 39 cases, the indication was a recurrent or residual aortic arch obstruction or coarctation in a patient aged 18 years or younger. This is our study cohort. The aorta was reconstructed by a direct anastomosis/autograft in 20 patients, patch in 17 patients, and interposition graft in 2 adolescents. In 32 procedures, cardiopulmonary bypass with whole body perfusion was employed, in 4, antegrade cerebral perfusion was employed, in 2, a left heart bypass was employed, and in 1 no cardiopulmonary bypass was used.

Results: Median (Q1, Q3) age at operation was 253 days (100, 2198 days), weight 7.5 kg (4.5, 17.8 kg). Median cardiopulmonary bypass time was 177 minutes (115, 219 minutes), crossclamp time 73 minutes (49, 102 minutes). Three infants died during the hospital stay: 1 with Williams syndrome, 1 with hypoplastic left heart syndrome, and 1 with heterotaxia. There was no death due to an arch complication. The main complications were 1 neurologic injury after postoperative resuscitation (Williams syndrome) and 1 permanent recurrent laryngeal nerve paralysis. During the follow-up period of median 8.1 years (2.6, 12 years) 1 re-reintervention on the aortic arch was necessary.

Conclusions: Sophisticated reoperations on the aortic arch could be performed safely. In children, the growth potential of all segments of the aorta could be sustainably preserved by avoiding interposition or extra-anatomic bypass grafts. (JTCVS Open 2024;19:215-22)



Angiography before/after repair of a recurrent AA stenosis in a patient with HLHS.

CENTRAL MESSAGE

The surgical anatomic repair of recurrent aortic arch stenosis can be performed safely and is a sustainable therapeutic approach.

PERSPECTIVE

The surgical treatment of a recurrent aortic arch stenosis is a highly challenging procedure. The 2 main therapeutic options are an anatomic repair on the 1 hand and an extra-anatomic aortic bypass graft on the other hand. This study confirms that the anatomic repair can be performed safely and seems to be a sustainable strategy in children and adolescents.

Aortic arch (AA) reconstruction in congenital heart surgery developed to a standardized and safe operation during the past decades. However, recurrent or residual stenosis still may occur.[1-3] Although transcatheter intervention is preferred in some cases, certain patients face limitations due to specific AA morphology or insufficient growth potential, particularly in children.[4,5] Anatomic repair techniques may be challenging because the approach to complete AA is never easy and even more complex in redo situations. Cardiopulmonary bypass strategies vary from deep hypothermic circulatory arrest to antegrade cerebral or whole body perfusion.[6-9] Some centers prefer extra-anatomic bypass grafts instead of the anatomic reconstruction.[10-12]

This retrospective single-center study aimed to assess mortality, complication rate, and mid-to long-term results of anatomic reconstruction for recurrent AA obstruction.

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Institutional review board approval: This retrospective study was approved by the ethics committee of the Medical Faculty at Johannes Kepler University Linz on May 17, 2023 (EK No.: 1269/2021).

Informed consent statement: Informed consent was waived because of the retrospective nature of the study and the use of pseudonymized clinical data for the analysis.

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Abbreviations and Acronyms
AA = aortic arch
REEEA = resection and extended end-to-end anastomosis

PATIENTS AND METHODS

Patients

This retrospective study was approved by the ethics committee of the medical faculty at Johannes Kepler University Linz on May 17, 2023 (EK No.: 1269/2021). Informed consent was waived because of the retrospective nature of the study and the use of pseudonymized clinical data for the analysis.

Statistical analyses were descriptive only and utilized Microsoft Excel for Mac version 16.78.3. Continuous variables were reported as medians and quartiles 1 and 3.

Between 1999 and 2022, 946 surgeries on the AA or isthmus were performed at the Children's Heart Center Linz, including 39 for recurrent or residual obstructions in patients aged 18 years or younger. This is our study cohort. We numbered the cases consecutively by the date of the AA reoperation (1 to 39). The patients' characteristics are provided in Table 1. According to the underlying disease, patients were divided into 3 anatomical subgroups: patients with isolated AA pathology, patients with biventricular circulation but an AA issue with additional cardiac defect, and univentricular patients. The AA reoperations in this univentricular cohort were performed at all stages of palliation, from 1 late conversion to 1 refenestration in a 12-year-old failing Fontan.

The indication for reoperation was an obstruction with a gradient of at least 20 mm Hg, in univentricular cases we do not accept any AA stenosis. The median (Q1, Q3) preoperative gradient was available in 35 patients: 34 mm Hg (25, 55 mm Hg) measured by sonography. In 13 cases (33%) a transcatheter dilatation, in 4 other (10%) patients stent implantations had been performed 2.8, 6.7, 7, and 14 years before the surgical reintervention.

Operative Technique

Since October 2003, we have performed all AA operations except an isolated coarctation repair using whole body perfusion and cardioplegic arrest (82% in this cohort). One arterial cannula was inserted into the innominate artery, either directly or by using a polytetrafluoroethylene prosthesis. In 30 patients, a second arterial cannula was placed into the descending aorta just above the diaphragm,[13] and in 2 patients with body weight >40 kg into a femoral artery. Antegrade cerebral perfusion was performed before this era. The approach for these 36 surgeries was a median sternotomy because this is our preferred approach also in borderline cases of distal AA obstruction.[3] In 3 cases, the restenosis was located at the isthmus region; therefore, the surgery was performed via a left thoracotomy. Twice, a left heart bypass (35 and 65 minutes) was used and once no bypass was used. In 1 infant (patient 31) the operation started with a median sternotomy and a thoracotomy was performed additionally because the end of the long interposition graft could only be reached by this approach.

We created a diagram that presents our strategy of decision making regarding the surgical approach (Figure 1): The 2 preferred methods are a resection and extended end-to-end anastomosis (REEEA) and an ascending-to-descending anastomosis. In the case of a patch reconstruction, we exclusively use patches curved in 2 planes out of a pulmonary homograft, an AA homograft, or a special vascular prosthesis (Figure 2).[14] If these techniques are not appropriate because of the lack of competent autologous material, we either perform a subclavian flap plasty or an aortic autograft (Figure 3, *A* and *B*). We try to completely avoid interposition grafts in children.

All different AA techniques used in our 39 cases of recurrent AA obstruction are presented in Figure 4. In the majority (19 patients), no patch was needed. In 1 12-year-old girl (patient 36) there was not enough competent autologous material at the distal AA, which was extremely long, narrow, and fragile. Therefore, an aortic autograft out of the ascending aorta was used (Figure 3, *A* and *B*).[15] In 16 cases, the AA was enlarged by a curved patch[14] together with an extended end-to-end anastomosis between the ascending aorta and AA (Figure 2), in patient 31 together with a subclavian flap plasty (Figure 5, *A* and *B*). In 2 adolescents (169 cm/60 kg and 165 cm/68 kg), the implantation of a 20-mm polyethylene terephthalate interposition graft had to be performed because of the lack of autologous material after removing a stenotic long interposition graft or stent. In 22 cases, additional cardiac procedures were necessary: 6 supravalvular and 4 subvalvular aortic stenosis repairs; 4 ventricular septal defect closures; 4 bidirectional Glenn operations; 3 pulmonary artery reconstructions; 2 systemic atrioventricular valve repairs; 2 right ventricle to pulmonary artery conduit exchanges; and each 1 Norwood procedure, Fontan operation, Fontan fenestration, Ross procedure, valve sparing aortic root replacement, aortic valve repair, and pacemaker procedure.

RESULTS

Postoperative Data and Early Mortality

Peri- and postoperative data are presented in Table 2. Thirty-six patients (92%) could be discharged from hospital, 3 patients died during the intensive care unit stay. Patient 5, a 6-month-old girl with heterotaxy syndrome, situs inversus, unbalanced atrioventricular canal, hypoplastic AA, and left atrial isomerism experienced a challenging recovery following her Norwood operation. Due to a gradient of 25 mm Hg at the distal AA with reduced ventricular function, she was reoperated by REEEA at age 6 months. Her ventricular function recovered slowly, but she ultimately died from a rhythm disorder on postoperative day 40. Patient 7 was a girl age 1 month who had a patch plasty performed at another center. In echocardiography we found a hypoplastic AA with a gradient of 30 mm Hg and severe mitral stenosis. We decided on a Norwood operation. Postoperatively, the saturation was low and catheterization showed pulmonary hypertension. This happened before the implementation of extracorporeal membrane oxygenation therapy at our center, and the baby died due to progredient ventricular failure on postoperative day 1. Patient 15 was a girl with Williams syndrome and status post REEEA. She underwent reoperation at age 53 days because of supravalvular aortic stenosis, hypoplastic AA, and hypoplastic pulmonary arteries. Unfortunately, she also had peripheral pulmonary stenoses and a persisting suprasystemic right ventricular pressure postoperatively. She died due to ventricular failure a few hours after the operation. In none of these mortality cases was a residual AA obstruction found, neither at pre- nor at postmortem examination.

Complications

The major complications in the intensive care unit were 3 cases of resuscitation. In 2 infants, this happened on the day

TABLE 1. Patients' characteristics and operative data

Subgroup	Isolated AA pathology	Biventricular complex	Univentricular	Total
Patients (female)	12 (6)	18 (8)	9 (4)	39 (18)
Age (days)	1535 (98, 4751)	352 (123, 1075)	124 (101, 184)	253 (100, 2198)
Weight (kg)	15 (5.5, 47.2)	9.2 (4.4, 13.5)	5.2 (4.1, 5.8)	7.5 (4.5, 17.8)
Time between first and second AA repair (d)	302 (92, 3436)	408 (81, 1221)	119 (96, 178)	224 (90, 1651)
Genetic syndrome				
Down	0 (0)	1 (6)	0 (0)	1 (3)
Williams	1 (8)	2 (11)	0 (0)	3 (8)
Heterotaxy	1 (8)	0 (0)	1 (11)	2 (5)
Duchenne	0 (0)	1 (6)	0 (0)	1 (3)
DiGeorge	0 (0)	2 (11)	0 (0)	2 (5)
Multiple malformations (no genetic examination)	0 (0)	1 (6)	0 (0)	1 (3)
Additional cardiac malformation	0	18 AS/LVOTO ± VSD (6) VSD (5) Williams syndrome/multiple vascular stenoses (2) Truncus arteriosus (1) CAVC (1) ccTGA/VSD (1) Taussig Bing (1) Mitral stenosis (1)	9 - HLHS (7) - Heterotaxy/hypoplastic left structures (1) - TA/TGA (1)	27
Interrupted AA	0 (0)	4 (22)	0 (0)	4 (10)
Primary approach				
Sternotomy	0 (0)	12 (67)	8 (89)	20 (51)
Left thoracotomy	12 (100)	6 (33)	1 (11)	19 (49)
Primary AA repair				
REEEA	6 (50)	12 (67)	0 (0)	18 (46)
Patch plasty	3 (25)	2 (11)	9 (100)	14 (35)
Interposition graft	3 (25)	0 (0)	0 (0)	3 (8)
Ascending-descending anastomosis	0 (0)	3 (17)	0 (0)	3 (8)
2 unknown AA reconstruction techniques + extra-anatomic bypass	0 (0)	1 (6)	0 (0)	1 (3)
Primary AA repair performed in our center	4 (33)	9 (50)	8 (89)	21 (54)
Stent in AA/isthmus	0 (0)	2 (11)	2 (22)	4 (10)

Values are presented as the median (Quartile 1, Quartile 3) or n (%). *AA*, Aortic arch; *AS*, aortic stenosis; *LVOTO*, left ventricular outflow tract obstruction; *VSD*, ventricular septal defect; *CAVC*, common atrio-ventricular canal; *ccTGA*, congenitally corrected transposition of the great arteries; *HLHS*, hypoplastic left heart syndrome; *TA*, tricuspid atresia; *TGA*, transposition of the great arteries; *R(E)EEA*, resection and (extended) end-to-end anastomosis.

of operation and in both a ventricular fibrillation could be terminated by defibrillation and volume substitution after a few minutes. The first patient (patient 9) ended up without any consequential sequelae, the second case concerned a boy with Williams syndrome (patient 4). Although no pathologies could be seen in his cerebral sonography examinations, the boy was still neurologically impaired at his final follow-up examination 19.5 years later. The third case of resuscitation was an infant with a multiple malformation syndrome. She happened to have a serious

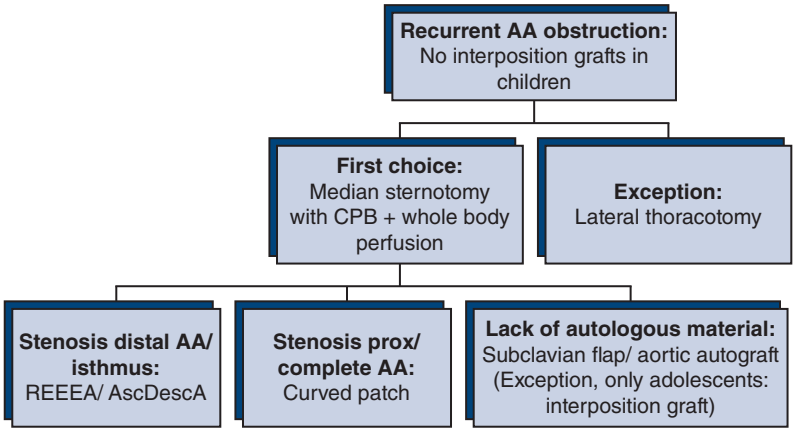

FIGURE 1. Decision making regarding the surgical approach at the Children's Heart Center Linz. *AA*, Aortic arch; *CPB*, cardiopulmonary bypass; *REEEA*, resection and extended end-to-end anastomosis; *AscDescA*, ascending-to-descending aortic anastomosis; *prox*, proximal.

bradycardia on postoperative day 9 after extubation. She was reintubated and ended up without any consequential damage; the second extubation was successful.

There was 1 postoperative bleeding and 1 hemothorax each with the indication for a rethoracotomy, both patients were in the complex biventricular group with further concurrent performed cardiac procedures. We had 1 sternal infection, 1 temporary unilateral phrenic nerve paralysis, and 4 patients with temporary postoperative stridor. A diagnostic examination regarding the function of the laryngeal nerves had not been performed routinely because the stridor had no serious clinical consequence. There was only 1 patient with a persistent unilateral laryngeal nerve paralysis at his final follow-up examination after 9 years; clinically he showed a slight hoarseness.

Long-term Outcome and Late Mortality

The follow-up period was defined as the interval between the AA reoperation and the final cardiac examination, including an echocardiography. At least 1 postoperative echocardiography of all 36 survivors was available. Therefore, the follow-up was complete and ranged from 10 days to 19.5 years (median 8.1 years; range, 2.6-12 years). There was no case of late mortality. One patient needed 2 unplanned re-reinterventions on the descending aorta (patient 31) (Figure 5, *A*): The AA could be repaired successfully, but the dissection of the very distal descending aorta could not be treated sufficiently by surgery. Therefore, 2 stents were implanted 7 days and 4.5 years later (Figure 5, *B*).

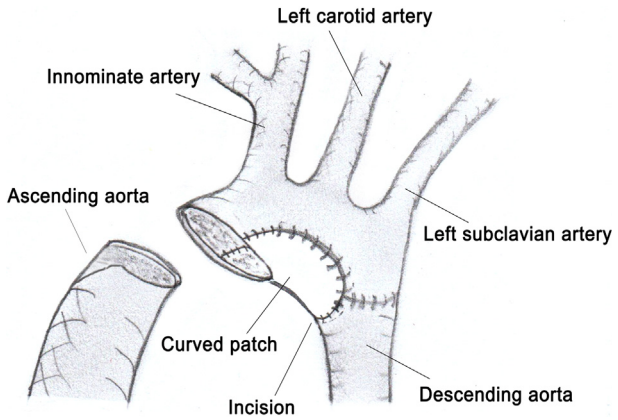

FIGURE 2. The most frequent performed technique for enlargement of the complete aortic arch (*AA*): resection of a re-coarctation, incision of the descending aorta, implantation of a curved patch, and extended end-to-end anastomosis between ascending aorta and AA.

In 32 out of 35 cases, no gradient and/or normal flow patterns at the AA and isthmus were described at final follow-up, 3 patients showed conspicuous results: Patient 38 still showed a gradient of 30 mm Hg at the proximal AA. The narrowest part of his former recurrent AA obstruction was found in a stent in his descending aorta. He underwent lateral thoracotomy for a stent removal and interposition graft (65 kg). Because there was no left heart hypertrophy at his final follow-up 9 months after this procedure, he currently stays under observation and will probably need an operation, including a right ventricle to pulmonary artery conduit exchange by sternotomy in the future. In patient 2, a slight recoarctation with a flow acceleration of 2.8 m/second, but normal flow patterns in the abdominal aorta, no aggravation and therefore no need for reintervention was described after 16 years of follow-up. In patient 4 with Williams syndrome (see Complications), a long-distance obstruction of the descending thoracic aorta with a gradient of 4.1 m/second was seen after 19.5 years. However, there is a normal abdominal aortic flow as well as an early deceleration and the gradient is probably overestimated. Additional diagnostic imaging will be necessary in the future, but his cardiologist and parents are reluctant because of his poor neurologic condition.

Unplanned reinterventions on other cardiac structures regarded the left pulmonary artery of 2 patients with hypoplastic left heart syndrome as well as the right or both pulmonary arteries after 2 Ross-Konno procedures and in

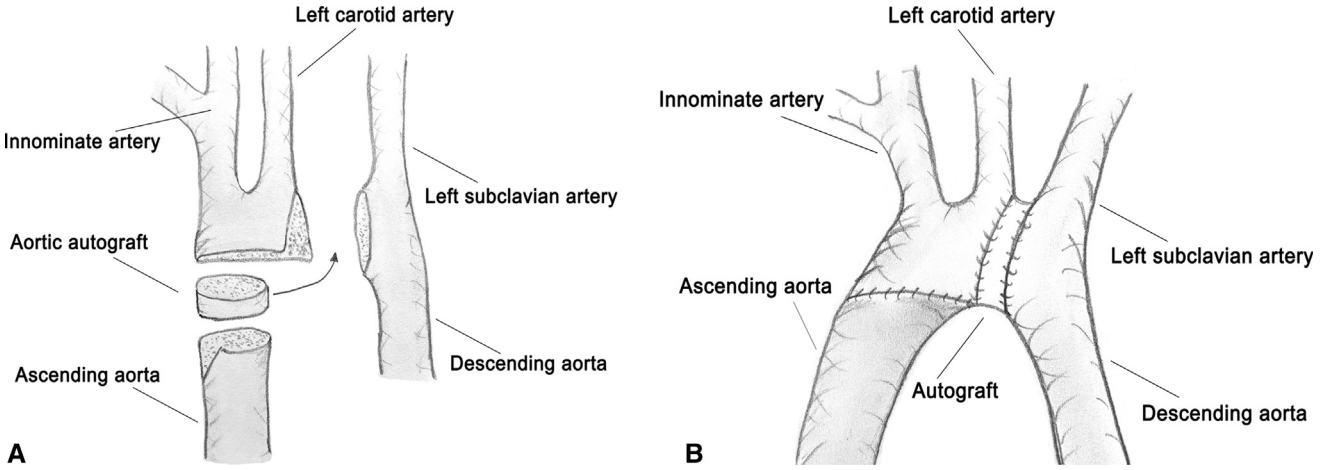

FIGURE 3. A and B, Use of an aortic autograft out of the ascending aorta for the reconstruction of the distal aortic arch.

1 further complex biventricular patient. There was no case with a left bronchial stenosis.

DISCUSSION

Anatomic Repair Versus Extra-Anatomic Aortic Bypass

The surgical treatment of a recurrent AA stenosis is a challenging procedure, and therefore alternatives as extra-anatomic aortic bypasses were invented. McKellar and colleagues[11] demonstrate very good intermediate-term results of an ascending-descending posterior pericardial bypass in a large cohort of 50 patients. However, this study only includes adults and adolescents using 21 ± 2 mm grafts. Kanter and colleagues[12] published their experience with ascending aorta-descending aorta bypass grafting in children, and they report no indication for reintervention during the follow-up period of mean 7.9 years using 10-mm grafts. Nevertheless, they conclude that the use of bypass grafts should be avoided whenever possible and an anatomic repair should be preferred. The main concerns, which are also mentioned in their discussion, are potentially difficult further operations, tethering of the aorta, and the development of aorto-esophageal fistulas.

In the current era of congenital cardiac surgery, the AA reconstruction developed to a routine operation, as can be seen on the total amount of 946 cases at our medium volume center. The different cardiopulmonary bypass techniques with the possibility of perfusing the brain and the lower parts of the body enable maximum safe conditions for even complex and long AA operations.[13] We are convinced

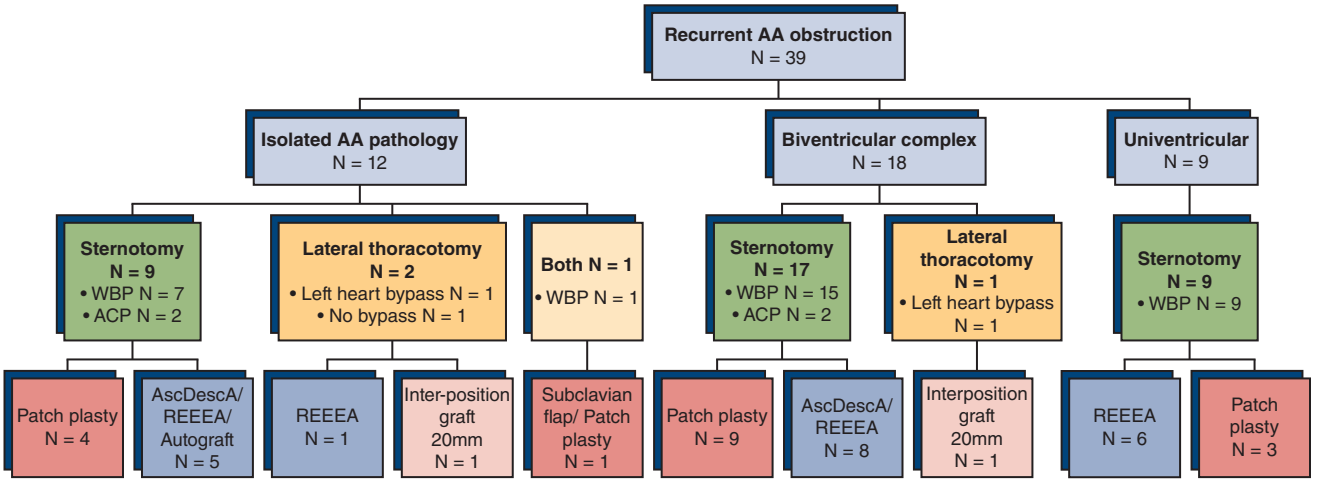

FIGURE 4. Surgical approach in the study cohort. *AA*, Aortic arch; *WPB*, whole body perfusion; *ACP*, antegrade cerebral perfusion; *AscDescA*, ascending-to-descending anastomosis; *REEEA*, resection and extended end-to-end anastomosis.

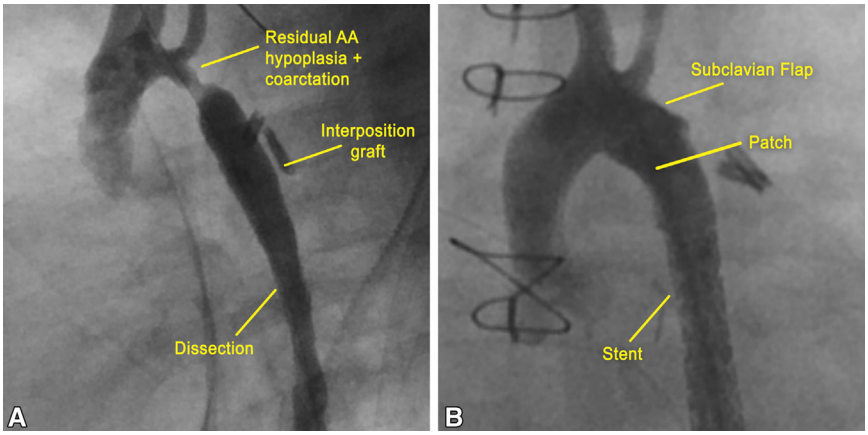

FIGURE 5. A and B, Only case of re-reintervention. A, Six-month-old girl's status after interposition graft. Residual stenosis at aortic arch (*AA*) and isthmus, dissection of the descending aorta. B, Stent implantation 7 days after reoperation.

that especially in young patients the surgical anatomic repair is the most sustainable strategy for recurrent or residual AA stenosis and coarctation (Figure 6). The different surgical techniques may be extremely challenging because the approach to the complete AA is never easy and even more complex in a redo situation. However, the chance of preserving the growth potential together with excellent results regarding complications, mortality, and reintervention rates of our study strengthens our opinion. Therefore, at the Children's Heart Center Linz no extra-anatomic bypass has ever been performed.

Surgical Approach

We now perform every reoperation on the AA through a median sternotomy on cardiopulmonary bypass during whole body perfusion. Exceptions are cases of recoarctation with implanted stents or interposition grafts that reach more distal parts of the descending aorta. Zoghbi and colleagues[5] published a large series of 63 patients, including adults, who underwent anatomic surgical repair for recurrent AA obstruction. Their approach in 52 cases (83%) was a left thoracotomy, and their outcome was impressive with 1 in-hospital death and 2 re-reinterventions. Our group[3]

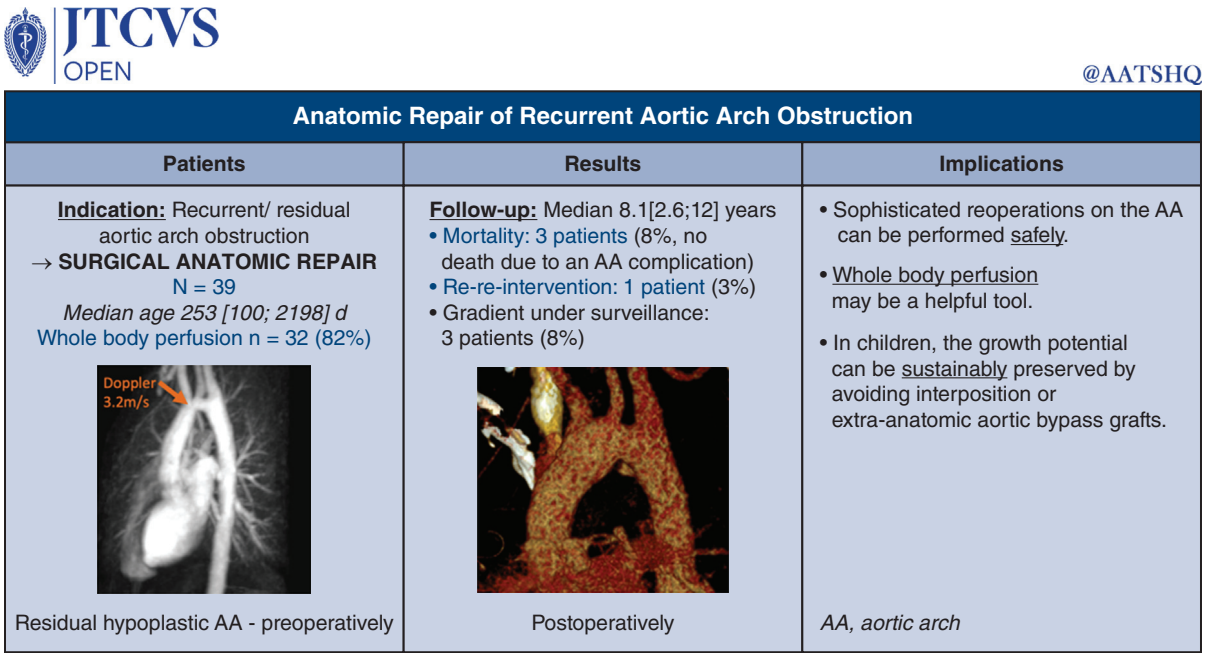

FIGURE 6. Graphical abstract. Magnetic resonance imaging. *Left*, After coarctation repair as a newborn infant, gradient aortic arch (*AA*) (echocardiography) 46 mm Hg. *Right*, Two years after reoperation.

TABLE 2. Peri- and postoperative data

Subgroup	Isolated AA pathology (n = 12)	Biventricular complex (n = 18)	Univentricular (n = 9)	Total (N = 39)
Bypass time (min), 38 patients	114 (88, 134) 11 patients	217 (179, 237) 18 patients	179 (131, 205) 9 patients	177 (115, 219)
Aortic crossclamp time (min) 36 patients	46 (30, 65) 10 patients	97 (91, 111) 17 patients	58 (45, 65) 9 patients	73 (49, 102)
In-hospital mortality	0 (0)	1 (6)	2 (22)	3 (8)
Stay on intensive care unit (d) 36 patients	3 (2, 6.5) 12 patients	9 (3, 11) 17 patients	14 (10, 16) 7 patients	7.5 (2, 12)
In hospital stay (d) 36 patients	9.5 (7.8, 12.5) 12 patients	14 (10, 23) 17 patients	21 (20,33.5) 7 patients	14 (9.8, 21.3)
Follow-up period (y) 36 patients	5.5 (2, 8.9) 12 patients	7.5 (3.4, 14.8) 17 patients	11.8 (8.9, 12.9) 7 patients	8.1 (2.6, 12)

Values are presented as median (Quartile 1, Quartile 3) or n (%). *AA*, Aortic arch.

identified an access through a left thoracotomy as the only risk factor for reintervention after AA or coarctation repair. We feel much safer with a median sternotomy because it allows addressing the entire AA. Furthermore, the time pressure decreases by using whole body perfusion, which we regard as essential during complex AA reinterventions.

Our strategy of avoiding interposition grafts or stents could not be put into practice in patients who were treated primarily by long interposition grafts or stents. This was the case in 2 adolescents weighing 60 and 68 kg due to limited availability of autologous material. However, in retrospect, using 20-mm interposition grafts in nearly fully grown patients did not pose significant problems, as evidenced by the absence of notable gradients during follow-up periods of 277 days and 11.7 years. Mery and colleagues[9] performed a very similar study with 48 patients aged 9 months (range, 22 days to 36 years) receiving a surgical repair of recurrent AA obstruction. Their results were excellent with 1 in-hospital mortality and 2 re-reinterventions. They advocate for reconstruction techniques utilizing autologous material or patch plasty, although interposition grafts were required in 2 cases.

In 1 special case from abroad, we tried very hard to restore an aorta with growth potential after removing a 2-cm-long interposition graft in a 6-month-old girl (patient 31) (Figure 5, *A* and *B*). We thought to be successful by performing a subclavian flap and a patch plasty, but could not treat the distal descending aorta surgically. This was the only case of re-reintervention in our center so far. However, we regard this hybrid strategy as superior to an interposition graft, given the potential for stent dilation. When the girl is older and hopefully nearly full grown, she probably will need an interposition graft.

Until now, we only used 1 aortic autograft (Figure 3, *A* and *B*) with no gradient after a follow-up of 645 days. Of course, this is not a long observation period. The use of an aortic autograft was already described by Metras and colleagues[15] for pulmonary artery reconstruction and it showed excellent long-term performance.

Several groups[9,16] describe the sliding arch aortoplasty as a further option for AA reconstruction without a patch, and they also used it for cases of reobstruction. Until now, we only performed this technique together with a curved patch (Figure 2), but it will be considered as a further option in future cases.

Transcatheter Reintervention

Transcatheter dilatation is an often-described option for a recoarctation,[17] and it is also standard in our center. A former study conducted by our group[3] describing a cohort of 139 patients who received an AA or isthmus repair in our center, showed 11 cases with recurrent AA obstruction, or coarctation necessitating reintervention. Eight of the reinterventions were performed by catheterization, 3 by surgery. In the current study, 33% of patients had a dilatation before the surgery. The exact date in several cases was missing because the transcatheter intervention had been performed in foreign centers. Considering the patients for whom the date was available, we saw that the surgical intervention was performed only days to weeks after the dilatation, demonstrating that the strategy was not successful in these patients.

Four patients of the current study were treated primarily by a stent 2.8 to 14 years preoperatively. Hence, this strategy seems to enable a substantial delay of the surgery. However, we regard an operation after stent implantation as much more difficult than a reoperation without a stent in a smaller patient, and the lack of autologous material after stent excision may be a problem (patient 38). Therefore, we do not see an indication for a stent implantation into the aorta of a child per se, and special cases should be discussed among cardiologists and surgeons before any reintervention (patient 31).

Mortality and Morbidity

Despite a lot of experience and innovative bypass techniques, serious complications may occur. In our series, there were 3 cases of in-hospital mortality and 1 case of cerebral

ischemia because of postoperative resuscitation. Two of these patients had complex univentricular conditions with Williams syndrome, in whom a high risk for acute heart failure after the correction of supravalvular aortic stenosis is described.[18,19] No coronary pathology was diagnosed in the 2 cases. However, this study should again raise awareness for the high perioperative risk in patients with Williams syndrome.

Limitations

The limitations of this study are the small number and the very heterogenous group of patients summarized as well as the lack of a control group. Further surveys with also longer follow-up periods will be necessary and useful.

CONCLUSIONS

Sophisticated reoperations on the AA could be performed safely by using whole body perfusion in the majority of procedures. The most serious complications and cases of death were associated with Williams syndrome or with complex univentricular conditions. There was 1 case of unplanned re-reintervention. The results concerning mortality, complication and re-reintervention rate confirm our strategy of preferring the anatomic repair to extra-anatomic bypass grafts even in complex cases of recurrent AA stenosis.

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

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