Autologous aortic arch reconstruction in isolated and combined cardiac lesions

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Summary

Objectives Various surgical strategies have been reported for the treatment of aortic coarctation with hypoplastic aortic arch, including simple resection and end-to-end anastomosis as well as various forms of patch augmentation. These techniques are limited by inadequate relief of arch obstruction and use of patch material predisposed to recurrent obstruction or aneurysm formation. We report our experience with autologous aortic arch reconstruction in isolated and combined lesions, a technique that relieves even complex forms of arch reconstruction without patch material.

Methods We retrospectively analyzed our institutional experience with autologous aortic arch reconstruction in isolated and combined cardiac lesions from November 2009 to December 2016. Study endpoints were procedural success, incidence of procedure-related complications, need for re-interventions, and survival.

Results In total, 54 patients underwent total autologous aortic arch reconstruction during the study period. Thereof, 13 (24%) had isolated arch obstruction and 41 (76%) had combined cardiac lesions. The majority of procedures were performed in the neonatal period (72%), median age was 8 days (range: 1 day to 4.3 years). Body weight ranged from 2.2 to 16.5 kg (median: 3.7 kg). There was one (1.9%) procedure-related early reoperation for bronchial obstruction. No repeat interventions (dilatation or re-operation) were observed. One patient with syndromic disease died on postoperative day 20 due to sepsis (1.9% in-hospital mortality rate). No late deaths were observed. Median follow-up was 23 months.

Conclusion Autologous aortic arch reconstruction is a safe and effective surgical technique for the treatment of aortic arch obstruction in isolated and complex cardiac lesions. It is associated with an extremely low re-intervention rate and a low overall complication rate.

Keywords Hypoplastic aortic arch · Congenital · Coarctation · End-to-side anastomosis · Aortic arch advancement

Introduction

Aortic arch hypoplasia is a frequent finding in patients with isolated aortic coarctation as well as in combined cardiac lesions. The success of surgery in patients with aortic arch hypoplasia is determined by the complete relief of obstruction, preservation of growth potential, and prevention of obstruction recurrence. Various surgical techniques have been developed for the treatment of aortic arch hypoplasia in patients with aortic coarctation and combined cardiac lesions over the
years. These include resection and end-to-end anastomosis [1, 2], subclavian flap technique [3], as well as various forms of patch augmentation [4, 5]. The latter are limited because they are predisposed to inadequate relief of obstruction and to aneurysm formation. Inadequate relief of arch obstruction has been related to a higher incidence of arterial hypertension as well as the development of relevant recurrent obstructions, necessitating re-interventions and limiting the long-term merits of surgery. In long-term studies, recurrent obstruction occurs in about 10% of cases [6, 7], with up to 16% [8] in the early decades. Aortic arch advancement with extended end-to-side anastomosis via a median sternotomy has been introduced to address the shortcomings of end-to-end anastomosis and patch augmentation [9–11]. This technique allows for complete relief of arch obstruction in isolated coarctation as well as in combined cardiac lesions without introduction of patch material into the aortic arch. Therefore, it facilitates our preferred approach of one-stage neonatal correction. We have exclusively used this technique in all patients presenting with severe aortic arch obstruction in isolated coarctation as well as in combined cardiac lesions since 2009. This retrospective analysis was conducted to evaluate our institutional experience with aortic arch advancement by end-to-side anastomosis. Study endpoints were procedural success, perioperative adverse events, and long-term survival as well as recurrence of arch obstruction.

Materials and methods

The Ethics Committee of the Medical University Vienna approved this study (EK Nr: 1582/2017). All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and with the Helsinki Declaration of 1975, as revised in 2008.

Study population

This is a retrospective analysis of all patients undergoing autologous aortic arch reconstruction in our center between November 2009 and December 2016. Patients were identified from the Viennese pediatric cardiac surgical database based on the ECHSA congenital database. Thereafter, a retrospective chart review was performed to obtain preoperative, operative, and inhospital data, as well as the most recent follow-up data from the outpatient clinic records.

Definition of hypoplastic aortic arch

Two-dimensional echocardiograms were used for preoperative assessment of cardiac structures and postoperative follow-up. For a first impression of the size of the aortic arch, we use the formula \( \text{diameter of transverse arch} = 1 + \text{weight (kg)} \) [10]. The aortic arch and isthmus region were measured and \( z \) scores calculated [12]. A \( z \) score of \( \leq 2 \) is defined as an hypoplastic aortic arch.

Surgical technique

Surgery was performed via a median sternotomy with cardiopulmonary bypass in all patients. For autologous aortic arch reconstruction and end-to-side anastomosis, the aortic arch, supra-aortic vessels, and the descending aorta were dissected free. Special emphasis was put on aggressive mobilization of the descending aorta as far distal as possible. Thereafter, the entire isthmic region was excised, including all ductal tissue, and the aortic arch and the distal ascending aorta were filleted open. The descending aorta was cut obliquely and a generous anastomosis between the incised distal ascending aorta, aortic arch, and descending aorta was constructed using Prolene 7-0. Perfusion strategies developed over time, from circulatory arrest to selective antegrade cerebral perfusion and additional distal aortic perfusion in more recent cases. Concomitant lesions were corrected with standard techniques.

Statistical analysis

Statistical analysis was performed with IBM SPSS Statistics for Windows, Version 23 (IBM Corp. released 2015; Armonk, NY, USA). Descriptive data were calculated as minimum, maximum, median, and interquartile ranges for non-normally distributed data. Normal distribution was ascertained with histograms (not shown). Categorical variables are presented as absolute numbers and percentages. The Poisson rate confidence interval was used to estimate risk of re-operation. For \( z \) scores of cardiac structures, especially the transverse aortic arch and isthmus, the Detroit data formula [12] and calculator were used with the Du Bois formula for body surface area.

Results

A total of 54 patients (male \( n = 37 \), 68.5%; female \( n = 17 \), 31.5%) underwent autologous aortic arch reconstruction during the study period. Patients with hypoplastic left heart syndrome undergoing Norwood-type procedures were excluded. Median age at surgery was 8 days (IQR: 5–45). The majority of patients were infants (<1 year; \( n = 52 \), 96%), with 39 (75%) neonates (<30 days). Two patients were older than 1 year. The oldest patient undergoing the procedure was 4.3 years.

Median weight at surgery was 3.7 kg (IQR: 3.1–4.3) with minimum of 2.2 kg and maximum of 16.5 kg. Median length at surgery was 51 cm (IQR: 50–55).

Thirteen (24%) patients had isolated aortic coarctation with hypoplastic aortic arch. The majority of patients (\( n = 41 \), 76%) presented with concomitant car-
Concomitant cardiac procedures are displayed in Table 1.

The calculated z score of the transverse aortic arch was a median of –4.4 (IQR: –5.1—–3.1), of the aortic isthmus, –6.0 (IQR: –8.0—–4.0). Calculation of the aortic arch with the formula \( \frac{\text{diameter of transverse arch}}{\text{weight (kg)}} \) [10] showed a median of –0.7 (IQR: –1.4—–0.2). Two patients were diagnosed with interrupted aortic arch.

Procedural success was 100%, with no conversions to a patch augmentation intraoperatively. One patient (1.9%) had to undergo early re-operation for left bronchial compression on postoperative day 12. The 30-day and in-hospital mortality rates were 1.9%. Postoperative complications occurred in 18 patients (33.3%; Table 2). These complications occurred primarily in patients after combined procedures (83%).

Median length of hospital stay was 18 days (IQR: 13–26). Correspondingly, length of ICU stay was 6 days (IQR: 4–9).

Median follow-up was 23 months, with a maximum follow-up of 6.3 years. No substantial residual gradients were measured postoperatively or during follow-up. No cases of late intervention in the catheter laboratory or surgical re-interventions were observed.

### Discussion

As shown in this retrospective study, aortic arch advancement can be successfully applied to a broad spectrum of congenital heart defects with aortic arch hypoplasia ranging from isolated coarctation with severely hypoplastic arch to complex combined cardiac lesions yielding excellent immediate and long-term outcomes.

Coarctation of the aorta with hypoplastic aortic arch is not exclusively an isolated problem. The majority of patients have concomitant lesions. For these combined defects, a median sternotomy offers the best approach for surgical repair. Furthermore, previous studies of a single-stage approach described good outcomes [13–15].

Total excision of ductal tissue is an important factor to lower the risk of re-coarctation [16]. Aneurysm creation was mostly described after patch plasty [6]. Aortic arch advancement via a median sternotomy with resection of the entire isthmic region, aggressive mobilization of the descending aorta, and end-to-side anastomosis has been developed to overcome the typical limitations of “traditional” end-to-end anastomosis, patch augmentation, and various forms of flap aortoplasty. The main benefit of this technique is the ability to create a generous and tension-free anastomosis that starts in the distal ascending aorta and sufficiently augments even severely hypoplastic aortic arches without the need for any form of patch material. Thereby, arch obstruction is effectively resolved and the growth potential of the arch is optimized while minimizing the risk of re-obstructions. Other techniques to avoid patch material are described in the literature as sliding plasty but are not compared with this study [17, 18].

We have systematically used this technique for aortic arch reconstruction in all patients presenting with isolated aortic coarctation and hypoplastic aortic arch (defined as \( \frac{\text{diameter of transverse arch}}{\text{weight (kg)}} \) [10]), as well as for patients with combined cardiac lesions and arch obstruction undergoing one-stage repair. In the latter group, the majority of patients fulfilled the criteria for arch hypoplasia; however, this technique was also used to facilitate one-stage repair in patients with dextro-Transposition of the Great Arteries (dTGA) complicated by aortic coarctation. Aortic arch advancement with extended end-to-side anastomosis could be successfully performed on all patients.

The majority of our patient cohort was younger than 12 months. Indeed, mostly neonates are in need of such an operation. However, in our study cohort there were two patients age 3.3 and 4.3 years, respectively. This indicates that this technique was not only
feasible in neonates and infants but also in larger children and in cases with recurrent arch obstruction. No intraoperative conversions to another technique or introduction of patch material was necessary even in patients with interruption of the aortic arch. This is in line with other groups that have demonstrated the reproducibility and versatility of this technique even in most complex arch obstructions.

It should be mentioned that this technique might change the natural geometry of the aortic arch. The development of a ‘‘gothic’’ morphology during growth is possible. Further investigations are needed to define whether this could lead to long-term complications. Contradictory statements exist in studies regarding the ‘‘gothic’’ arch related to these issues [19, 20].

Postoperative complications after coarctation repair occur in 36% of cases according to an evaluation of the STS database. The likelihood of complications is higher in a subgroup of patients with concomitant lesions [21]. This group is comparable with our cohort. The overall perioperative complication rate in the present study was low. Importantly, no cerebral complications related to circulatory arrest or antegrade cerebral perfusion were observed. Note-worthy, the incidence of perioperative renal failure was also low. Two distinct complications that clearly relate to the surgical technique were observed. One patient developed relevant left bronchial compression in the immediate postoperative period and had to undergo early reoperation. Left bronchial compression in this case was caused by a combination of insufficient mobilization of the descending aorta and aortic arch together with an end-to-side anastomosis, created to proximal in the ascending aorta. This problem occurred early in this series and could certainly be avoided. Furthermore, we observed transient left-sided vocal cord paralysis in 9.3% of patients, which can be explained best by intraoperative traction injury to the left recurrent laryngeal nerve. This is a known complication of operations of a coarctation of the aorta. Although it has been reported in up to 38% of cases in the literature [22], there is low residual dysfunction. This complication may be prevented by avoiding any intraoperative traction in the area of the recurrent laryngeal nerve.

The major controversy in the comparison of aortic arch advancement with extended end-to-side anastomosis and the repair via a lateral thoracotomy is the necessity to use cardiopulmonary bypass. This can be maintained in combination with brief circulatory arrest, antegrade cerebral perfusion, or combined antegrade cerebral plus descending aortic perfusion. This significant extension of the procedure has to be weighed against the risk of re-obstruction, especially in patients with isolated coarctation and hypoplastic arch. It has been advocated that the majority of patients with aortic coarctation and hypoplastic aortic arch can be operated on via a lateral thoracotomy and that residual arch obstruction will resolve spontaneously with growth after resection of the coarctation. Our data clearly demonstrate that the outcome with the pursued approach is excellent. The complication rates observed are exceptionally low as is re-

![Fig. 1 Kaplan–Meier curve for freedom from re-intervention](image-url)
currence of obstruction. This has to be put in context with the ever-increasing evidence that residual obstruction is associated with worse long-term outcomes and higher incidences of recurrent obstruction within the first 10 years of surgery.

**Limitations**

The present study is affected by the typical limitations of a retrospective study and lacks a control group.

Further limitations should be mentioned: Various methods of calculation and assessment of the aortic arch exist [23]. Hence, there is no unique classification for aortic arch hypoplasia. We rely on the simple formula of \( \text{diameter of transverse arch should be} \geq 1 + \text{weight (kg)} \times 10 \), which has also been applied by others in comparable studies. In addition, monitoring for re-coarctation was performed with ultrasound only. In all patients, the pressure gradient between the upper and lower body was denoted to be unessential slight, if any, postoperatively and during follow-up. However, no specific blood pressure data were obtained.

**Conclusion**

In conclusion, autologous aortic arch reconstruction with end-to-side anastomosis is a safe and effective surgical technique for the relief of aortic arch obstruction in isolated and combined cardiac lesions. It is associated with an extremely low re-intervention rate (see Fig. 1) and can be performed with a low overall complication rate.

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**Compliance with ethical guidelines**

**Conflict of interest** C. Herbst, G. Laufer, S. Greil, E. Kitzmueller, E. Base, R. Vargha, and D. Zimpfer declare that they have no competing interests.

**Ethical standards** Informed consent was obtained from all patients for being included in the hospital data registry. As this is a retrospective study, no separate informed consent was needed in regard to this study.

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