Management of congenital tracheal stenosis in infancy

Tsvetomir Loukanov*, Christian Sebeninga, Matthias Gorenfloa, Siegfried Haglab
aDepartment of Cardiac Surgery, University of Heidelberg, INF 110, D-69120 Heidelberg, Germany
bDepartment of Pediatric Cardiology, University of Heidelberg, INF 110, D-69120 Heidelberg, Germany

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We read with great interest the article of Anton-Pacheco et al. [1] in which they analyzed 19 cases with congenital tracheal stenosis. This single center study consisted of different groups of patients, classified to the severity and length of the tracheal stenosis. The authors described three different surgical techniques with an overall mortality rate of 21%. The rate of mortality in Anton-Pacheco’s series exceeds clearly all contemporary standards published in the literature in the last decade [2] (early mortality 6% and late mortality 12%). The authors reported excellent results with slide tracheoplasty in 7 patients but very poor clinical outcome with costal cartilage tracheoplasty. Cardiopulmonary bypass (CPB) was used in all patients with slide tracheoplasty but only occasionally in cases using the cartilage tracheoplasty and in cases presenting with short segment stenosis.

According to our experience based on 37 patients with tracheal pathology, isolated or associated with congenital heart malformations the use of CPB in the surgical management of these patients is very useful and represents an effective tool. In our hands the survival rate was 97% despite of the associated cardiovascular malformations, higher percentage of redo procedures and the complexity of surgical interventions. We attempted surgical treatment of tracheal pathology with simultaneous repair of congenital cardiac lesions in most of the cases [3]. The crucial part of our surgical technique regarding the tracheal part of the operation was careful dissection and mobilization of the trachea, main bronchi, pulmonary arteries and veins in order to obtain tension-free tracheal anastomosis.

In our experience the key element in the surgical approach for such patients represents the use of CPB which allowed extensive mobilization of the whole tracheobronchial tree and resection of the trachea with end-to-end tension free anastomotic reconstruction. The use of CPB avoids any additional tension when placing or tying the sutures at the tracheal anastomotic site. The use of CPB allows mobilization of longer (above 30%) segments of the trachea which are then feasible for resection and reconstruction with end-to-end anastomosis.

We emphasize also on the intraoperative use of endoscopic monitoring of the airways. It permits careful exploration for identifying residual tracheal stenosis and facilitates clearing of secretions at the end of operation.

We are strongly convinced that the use of CPB and monitoring of the surgical procedure by intraoperative bronchoscopy are the cornerstones in the successful management of patients with congenital tracheal stenosis.

References


The authors of the original paper [1] were invited to comment on this Letter to the Editor but declined the offer.

* Corresponding author. Tel.: +49 6221 56 6272; fax: +49 6221 56 5919. E-mail address: Tsvetomir.Loukanov@med.uni-heidelberg.de (T. Loukanov).

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Letter to the Editor

When should we replace the ascending aorta in Marfan syndrome?

Mehmet Ates*
Department of Cardiovascular Surgery, Prof. Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Center, Istanbul, Turkey

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I read with interest the article entitled ‘Chronic dissection of the ascending aorta: surgical results during..."
a 20-year period (previous surgery excluded) by Jault et al. [1]. They have analyzed the surgical aspects of aortic disease in a 20-year period. I congratulate them for this article and would like to add few comments. Replacement of the ascending aorta is the most frequently performed procedure for thoracic aortic pathology. Current indications for replacement of the ascending aorta and aortic root may be divided into two broad categories: mandatory indications and elective indications. Mandatory indications are usually urgent situations involving acute dissection of the ascending aorta and related pathology. Elective indications are relative indications, as these operations are generally prophylactic in nature, aiming to prevent progression of aortic insufficiency and rupture or dissection of the aorta in Marfan’s syndrome related pathology in the presence of degenerative dilatation of the ascending aorta [2].

Our question is when we should replace the ascending aorta in Marfan syndrome. The classical attitudes are as follows: for degenerative aneurysms, 6 cm; for non-Marfan annulo-aortic ectasia, 5.5 cm and for Marfan patients, 5 cm [3]. Recently these diameters are decreased by cardiac surgeons as follows: for degenerative aneurysm; 5 cm and for Marfan patients; 4.5 cm [4]. Legget et al. [5] showed that in Marfan’s syndrome, an initial aortic ratio (measured diameter/predicted diameter) at the sinuses of 1.3 increased the relative risk of rupture, dissection or operation for enlarged diameter by 2.7, and a >5% annual increase in the aortic ratio increased these risks by a factor of 4.1. They suggest that patients with dimensions smaller than these parameters could be observed with some degree of comfort. In our institute, if aortic diameter is more than 4.5 cm, we are replacing the ascending aorta in Marfan syndrome and also if it is possible, we are saving aortic valve.

Reply to the Letter to the Editor

Frédérique Jault*, Beltran Levy Praschker
Department of Thoracic and Cardiovascular Surgery, Professor Gandjbakhch Service, University of Paris VI Pierre et Marie Curie, GH Pitié-Salpêtrière, Assistance Publique Hopitaux de Paris, 47-83, boulevard de l’Hôpital, 75651 Paris Cedex 13, France

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We thank Ates [1] for his remarks and interest in our work [2]. We published 12 years ago [3], a report on 339 patients who underwent surgery of the ascending aorta, including all chronic pathologies and redo patients; in our recent data [2], we focused on patients with chronic dissection of the ascending aorta who have never been operated before; the majority had severe aortic insufficiency. Clinical presentation, operative techniques and results are different according to the different pathologies, and of course are quite different in acute aortic disease, but it was not our subject.

In our report [2], the extent of the dissection is the only risk factor for operative and late mortality; 15.5% of patients had Marfan disease and 23% annulo aortic disease.

In Marfan disease, when there is no dissection, as the question raised by Ates [1], when the ascending aorta should be replaced? Aortic diameter should be measured at the sinuses of Valsalva, and related to normal values based on age and body surface area [4], especially in paediatric patients. Surgery is recommended by most surgeons when the aorta is >4.5 cm (in adults), when rapid growth of the aortic diameter occurs (>1 cm/year), when there is a family history of aortic dissection, when there is a greater than mild aortic insufficiency [5]. Some surgeons consider prophylactic aortic root replacement; this is not validated, but there is a tendency to operate at smaller diameters, when aortic insufficiency is mild or inexistent.

Composite valve graft repair is used since 1968; the long-term results are good [3]. Valve-sparing aortic root replacement is used more recently [5]. Several techniques such as Yacoub procedure (remodeling technique) and David procedure (reimplantation technique) are used.

The aortic insufficiency, when present, can be corrected, but for some surgeons, structural anomalies of valve cusps and severe aortic insufficiency are a contra-indication for these procedures. The main problem is in fact the durability of the preserved aortic valve. Fleischer had shown fragmentation and scarcity of fibrillin in the excised aortic leaflets in Marfan disease. Most of the problems emerge in the 5–10 years time; 25% of the patients at 10 years had 3+ to 4+ aortic insufficiency, especially in paediatric patients.

Patients with Marfan disease selecting a valve-sparing procedure must be warned of possible reoperation in the future.

Last, arch and descending aorta are sites for later aneurysms and dissection.

References


* Corresponding author. Address: Merdivenkoy Mah., Sairarsi Cad., Emincinapasa Sok., No: 6/24 Goztepe-Kadikoy, 34732 Istanbul, Turkey. Tel.: +90 216 360 0272; fax: +90 216 337 9719.

E-mail address: drmates@yahoo.com.

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