Aortic Valve Atresia or Severe Hypoplasia and Ventricular Septal Defect: Surgical Strategies for Biventricular Repair and Mid-term Results

Atresia valvular aórtica o hipoplasia grave y comunicación interventricular: estrategias de corrección biventricular y resultados a medio plazo

To the Editor,

Aortic valve atresia, or severe hypoplasia associated with ventricular septal defect and normal-sized ventricles, is an exceptional entity in neonates. Biventricular correction is feasible in a single intervention (described by Yasui in 1987)\(^1\) or in 2 stages, using a Norwood procedure followed by a Rastelli operation in order to subsequently establish biventricular physiology (successfully used in 1981).\(^2\) This approach involves reconstructing the ascending aorta by using the Damus-Kaye-Stansel technique (Figure A) and closing the septal defect and the conduit between the right ventricle and the pulmonary artery (Figure B), sometimes combined with reconstruction of the aortic arch. We present the results of the medium-term follow-up in our series.

Between 1990 and 2013, 27 patients (mean age, 19 [2-112] days; mean weight, 3.25 [2.1-5.2] kg) with this diagnosis (aortic valve atresia in 18 and severe hypoplasia in 9, defined as ring diameter < 3 mm) underwent biventricular repair (interrupted aortic arch in 14 and aortic coarctation in 7). Primary repair (Yasui surgery)\(^3\) was performed in 19 patients (70.4%), and the 2-stage approach in 8 (29.6%), using the Norwood\(^4\) technique followed by a Rastelli operation at 9.6 (3.3-29.2) months.

The inclusion criteria (based on 2-dimensional echocardiography) for choosing this repair technique were lack of fibroelastosis in the left ventricle, good ventricular function, mitral ring z-score > -2, left ventricle quotient (long axis) > 0.8, left ventricle ≥ 20 mL/m\(^2\) and nonrestrictive ventricular septal defect. The staged approach was implemented as standard in 2008.

Data were extracted from our database (HeartSuite, Systeria, Inc, Glasgow, United Kingdom) and from medical records. The analysis was performed using R software v3.0.2 (R Foundation for Statistical Computing, Vienna, Austria). The Kaplan-Meier (log-rank test) was used for the survival analysis. A P-value of < .05 was considered statistically significant.

Overall 30-day mortality was 25.9% (n = 7), and operative mortality was 7.4% (n = 2, both following Yasui\(^1\) surgery with

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hours. With the aim of correcting the valve disorder, we discussed the case with the surgeons, who decided that the risk of surgery would be too high (Society of Thoracic Surgeons [STS] score: mortality, 6.7%; logistic EuroScore, 29.1%).

Due to the improbability of the patient recovering well if the valve disorder was not corrected, we opted for mitral repair with the MitaClip\(^6\) system (Abbott Vascular). A transesophageal echocardiography was performed, which showed severe MR with regurgitation in the A2-P2 scallops, although with a certain degree of medial involvement (A3-P3) (Figures 1E and F).

The procedure was performed under general anesthesia and under transesophageal echocardiographic guidance. After grasping both leaflets in the A2-P2 position, regurgitation reduced noticeably, although residual moderate MR persisted in the area lateral to the implanted clip (Figure 2A). We therefore implanted another clip, lateral to the former, managing to reduce MR to < 1/4 (Figures 2B and C).

The course was favorable thereafter, with complete disappearance of symptoms and signs of heart failure. A check-up echocardiogram 4 days later showed mild residual MR (Figure 2C). The patient was discharged from hospital and is currently New York Heart Association class I.

Acute MR in a context of infarction with ST elevation due to papillary muscle dysfunction is considered to be a mechanical complication that usually causes the patient’s clinical worsening and leads to pulmonary edema and sometimes cardiogenic shock. In this scenario, mitral surgery is considered to be the treatment of choice. However, this kind of surgery is associated with high mortality\(^1\) due to the high risk profile of patients, which is why surgeons sometimes decide against it. This is the context in which transcatheter valvular treatment techniques are gaining respect. The MitaClip\(^6\) is the only device for treating mitral valves that is widely used on a clinical level, and mainly in Europe. Recent studies show that the use of this device is safe and effective, achieving functional class improvement in 80% of cases and a persistent reduction in MR after one year.\(^2\)\(^-\)^\(^4\) However, the information on treating patients with acute MR is scarce.\(^5\) As this is a phenomenon of recent onset and of functional etiology, the leaflets present characteristics of abundant tissue and coaptation surface that are usually suitable for implanting a clip. Correction of MR leads to rapid clinical recovery, as the volumetric overload is effectively corrected, but there are few data on the effect of the clip on ventricular remodeling after infarction. More studies, with appropriate imaging follow-up, are therefore necessary.

This case shows that mitral repair with the MitaClip\(^6\) device is a safe and effective technique for correcting acute MR following infarction in patients at high surgical risk. This fact could extend the indications for use of the device to include this patient population.

Miguel Rodríguez-Santamarta, Rodrigo Estévez-Loureiro,* Javier Gualís, David Alonso, Armando Pérez de Prado, and Felipe Fernández-Vázquez.

Departamento de Cardiología, Complejo Asistencial Universitario de León, León, Spain

*Corresponding author:
E-mail address: roiestevez@hotmail.com (R. Estévez-Loureiro).
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failure of extracorporeal circulation during the interval before
the implantation of a longer-term circulatory support tech-
nique). The remaining causes of early death were of non-cardiac
origin. There were no deaths between the stages of the 2-stage
approach. Mortality during follow-up (mean [standard devia-
tion], 8.4 [5.3] [0.9 to 23] years) was 10.5% (n = 2, both following
primary correction: sudden death of undetermined origin,
episodes of ventricular tachycardia, and irreversible cardiac
arrest). Of the 20 survivors, 9 (45%) required intervention (arch
stenosis or recoarctation in 5, and conduit stenosis in 9), and
9 underwent surgery again for conduit replacement (pacemaker
implantation in 1 patient after obstructive resection of the left
outflow tract). Of the initial survivors (n = 20) 95%, 85%, 50%
and 35% had not required reintervention after 1, 2, 5, and
10 years, and 70%, 65%, and 55% were without percutaneous
procedures after 1, 2, and 5 years. The incidence of reinterven-
tion and interventionism in each group according to the surgical
technique used is shown in the Table. Overall survival during
follow-up (n = 20) was 95% at 10 years.

Primary correction and 2-stage surgery are 2 options with
very similar results for biventricular correction in this context,3,4
representing a safe and effective strategy with excellent medium-
term survival, although with a high rate of reintervention/
interventionism.

Yasui surgery1 (a high-risk procedure for neonates, who are
occasionally underweight), avoids provision al univentricular
physiology and allows complete correction. The Norwood
technique5 avoids the initial complexity of corrective surgery,
which involves 2 normal ventricles contributing to cardiac output,
and allows a period of growth prior to final biventricular
correction. This may facilitate better selection of candidates, as
there may be some patients who will benefit from a
univentricular physiology and who could not otherwise have
been identified, which is especially important in the case of
impaired left ventricular function.

A study by the Congenital Heart Surgeons’ Society Data Center
(Toronto) showed that primary biventricular repair in neonates
in cases of critical aortic stenosis led to a high reintervention
rate (50% at 3 years). This resulted in a 30-day mortality of 60%,
which could reflect an inadequate initial indication,5,6
possibly due to a belief that early septation must be achieved. However,
the rationale for choosing one or other surgical option remains
controversial and will continue to be debated.

María-Teresa González-López, a, * Timothy J. Jones, a John Stickley, b
David J. Barron, a Natasha Khan, a and William J. Brawn a

aPediatric Cardiac Surgery Department, Heart Unit,
Birmingham Children’s Hospital, Birmingham, United Kingdom
bStatistical and Investigation Department, Heart Unit,
Birmingham Children’s Hospital, Birmingham, United Kingdom

*Corresponding author:
E-mail address: draglezlopez@hotmail.com
(M.-T. González-López).
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Fate of Original Articles Rejected by Revista Española de Cardiología

Destino de los artículos originales rechazados en REVISTA ESPANOLA DE CARDIOLOGIA

To the Editor,

A crucial phase in the editorial process of a scientific article is its review by a panel of peers, who analyze its quality and provide constructive criticism to enable the authors to improve the manuscript. Their opinions aid the editor in making a decision on the article’s suitability for publication. The objective of this study was to analyze the final destination of original articles rejected by Revista Española de Cardiología (Rev Esp Cardiol).

We accessed the database of the online submission and review system of Revista Española de Cardiología to compile a list of the 650 original articles rejected over the 4-year period between 1 January 2007 and 31 December 2010. The following data were collected for each article: manuscript reference number, title, authors, type of article, rejection date, article version (number), language, and country of origin (or, in the absence of the latter datum, that of the corresponding author). The resulting list was distributed equally among 8 members of the editorial board of Rev Esp Cardiol, who carried out the corresponding searches in the literature published up to March 2014.

The literature searches were performed in accordance with the following indications: a) to access the PubMed database and copy the title of the article in English; b) if the search failed to yield results, to use the surname and initial(s) of the corresponding author of the article; c) if there were still no results with the preceding measure, and the corresponding and first authors were not the same person, to repeat the search using the surname and initial(s) of the first author. When the search produced results that coincided with a published article, the following information was recorded in a new database: journal, the year the paper edition was printed, date accepted, impact factor assigned to the journal by Journal Citation Reports for the year the article was published, type of article, language, and names of the first and corresponding authors. A positive result was defined as that in which the methodology of the abstract of the article coincided with that of the publication found, regardless of the type of article finally published, except in the case of communications and posters presented at congresses, which were not taken into account. In a second phase, 2 members of the board reviewed the articles one-by-one to fill in all the empty or incomplete boxes. Finally, the information for those articles not producing results in the PubMed search was introduced into Google.com, following steps a, b, and c, as described above.

During the study period, Rev Esp Cardiol rejected 650 original articles (102 in 2007, 142 in 2008, 192 in 2009, and 214 in 2010). Of the 650 rejected original articles, 96% were in the original version, 3.4% had been revised for the first time, and 0.6% had been revised for the second time. Regarding the language of manuscript submission, 80.9% were in Spanish and 19.1% in English. The rejected articles, in descending order of frequency, were sent from Spain, Argentina, Mexico, China, Brazil, Cuba, and Chile.

As of March 2014, of these 650 articles, a total of 287 (44.1% of all rejected papers) have been published. They have been distributed in different article types (in some cases, not comparable from one journal to another), but most (79.4%) retained the format as the initial version. Sixty-five with five percent of these articles were published within 2 years after their rejection by Rev Esp Cardiol. They appeared in 140 different journals, and 36% of the manuscripts were concentrated in 10 journals (9 of which publish in Spanish, whereas the remaining journal publishes only in English). The 5 journals in which these articles most frequently appeared were Medicina Clínica (32 articles), Revista Clínica Española (13 articles), Revista Médica de Chile (10 articles), International Journal of Cardiology (10 articles), and Atención

Of the 228 rejected articles received in Spanish for consideration by Rev Esp Cardiol, 166 (65.6%) were published in Spanish, 48 were published exclusively in English, 11 in Spanish and English, 2 in English and Portuguese, and 1 exclusively in Portuguese.

Of the 59 rejected articles received in English for consideration by Rev Esp Cardiol, 49 (83%) were published in English, 4 were published exclusively in Spanish, 4 in English and Portuguese, 1 exclusively in Portuguese, and 1 in Arabic and English.

Figure. Publication language of the 287 rejected articles subsequently published in other journals.