Atrioventricular Septal Defect Prognosis for Patients With Down Syndrome

Marta Unolt · Carolina Putotto · Dario Marino

Received: 13 July 2012 / Accepted: 7 August 2012 / Published online: 25 August 2012
© Springer Science+Business Media, LLC 2012

We read with interest the article by Patel et al. [8] entitled Early Outcomes and Prognostic Factors for Left Atrioventricular Valve Replacement after Primary Atrioventricular Septal Defect Repair in a recent issue of your journal. In their cohort of patients with previously repaired atrioventricular septal defect (AVSD), the authors identified Down syndrome (DS) as an important predictor of in-hospital mortality after left atrioventricular valve (LAVV) replacement. Surprisingly, the study contains no possible explanations for these data.

Moreover the authors wrote that “the association between DS and survival after surgical interventions for AVSD remains unclear” [8]. In our opinion this sentence is not accurate and in this form could be misleading.

In the recent literature, only one study of a cohort from the period 1958 to 1997 reported that infants with complete AVSD and DS at surgical correction had a higher mortality rate than infants without DS [9]. However, as previously noted [7], the reported mortality difference was primarily in the earlier years of the study [9] when infants with AVSD and DS probably received unequal cardiac care [7]. All recent contributions have clearly demonstrated that with the current level of care, DS is not a risk factor for primary surgical correction of complete AVSD [1–7]. Moreover, some studies have even suggested that DS in patients with complete AVSD may be considered a protective factor for surgical correction [1, 3, 5] and for long-term follow-up experience [1, 2] due to the more favorable cardiac anatomy. These are fundamental and established data for correct counseling in relation to fetuses, neonates, and children with DS and AVSD.

For patients with DS, the higher mortality rate at valve replacement after primary correction of AVSD is new and interesting information [8], and this topic deserves further studies, additional data, and adequate explanations.

References

1. Al-Hay AA, MacNeill SJ, Yacoub M, Shore DF, Shinebourne EA (2003) Complete atrioventricular septal defect, Down syndrome, and surgical outcome: risk factors. *Ann Thorac Surg* 75:412–421
2. Boening A, Scheewe J, Heine K, Hedderich J, Regensburger D, Kramer HH, Cremer J (2002) Long-term results after surgical correction of atrioventricular septal defects. *Eur J Cardiothorac Surg* 22:167–173
3. Formigari R, Di Donato RM, Gargiulo G, Di Carlo D, Feltri C, Picchio FM, Marino B (2004) Better surgical prognosis for patients with complete atrioventricular septal defect and Down's syndrome. *Ann Thorac Surg* 78:666–672
4. Fudge JC Jr, Li S, Jagers J, O'Brien SM, Peterson ED, Jacobs JP, Welke KF, Jacobs ML, Li JS, Pasquali SK (2010) Congenital heart surgery outcomes in Down syndrome: analysis of a national clinical database. *Pediatrics* 126:315–322
5. Lacour-Gayet F, Bonnet N, Piot D, Serraf A, Sousa-Uva M, Belli E, Roussin R, Bruniaux J, Planché C (1997) Surgical management of atrioventricular septal defects with normal caryotype. *Eur J Cardiothorac Surg* 11:466–472
6. Lange R, Guenther T, Busch R, Hess J, Schreiber C (2007) The presence of Down syndrome is not a risk factor in complete atrioventricular septal defect repair. *J Thorac Cardiovasc Surg* 134:304–310
7. Miller A, Siffel C, Lu C, Riehle-Colarusso T, Frías JL, Correa A (2012) Long-term survival of infants with atrioventricular septal defects. *J Pediatr* 156:994–1000
8. Patel SS, Burns TL, Kochilas L (2012) Early outcomes and prognostic factors for left atrioventricular valve reoperation after primary atrioventricular septal defect repair. *Pediatr Cardiol* 33:129–140
9. Reller MD, Morris CD (1998) Is Down syndrome a risk factor for poor outcome after repair of congenital heart defects? *J Pediatr* 132:738–741

M. Unolt (✉) · C. Putotto · D. Marino
Department of Pediatrics, “La Sapienza” University,
Viale Regina Elena, 324, 00161 Rome, Italy
e-mail: marta_unolt@hotmail.it