LETTERS TO THE EDITOR

Transposition of the great arteries

To the Editor:

I read with interest the report by Houyel and associates[1] on transposition of the great arteries {S,D,L}, a complex not so newly recognized as the authors suggested.

As reported by the authors and some others previously,[2-5] transposition of the great arteries with levomalposed great arteries (L-TGA) is a rare anomaly (0.3% of a large series of specimens with congenital heart malformations and 4% of cases of anatomically proven complete TGA[2]). Some findings of L-TGA include a high frequency of certain associated anomalies, specifically, abnormalities of the position of the cardiac apex, juxtaposition of the atrial appendages, anomalies of the atrioventricular valves, ventricular septal defects (VSDs), pulmonary outflow tract obstruction, abnormal infundibular pattern,[2-5] and ventricular malrotation.[4, 5] It shares these findings with other closely related anomalies characterized by a levoposed aorta and D-loop (noninverted) ventricles, including some types of double-outlet right and left ventricles and some types of anatomically corrected malposition. Years ago my colleagues and I[2-5] suggested that a syndrome (then called *aortic levoposition without ventricular inversion*) existed that included all these anomalies and could be produced by similar morphogenetic faults. This raised the possibility of the existence of a common spectrum of development wherein minor differences in the alignment of conus and ventricles could produce, for example, L-double-outlet right ventricle with subpulmonary VSD, L-TGA, or L-double-outlet left ventricle with subaortic VSD.

The report by Houyel and associates and our previous reports, as well as the report by Lincoln and associates,[6] show similar findings on the very high frequency of VSDs (with a peculiar anatomy that deserves emphasis, most being of malalignment type and some caused by combined malalignment and infundibular septal deficiency) and conoventricular malalignment. A difference between our series, however, is the frequent presence of a bilateral conus in our cases whereas Houyel's group found only one case.

Pulmonary outflow tract stenosis was more frequent in our cases, whereas right ventricular hypoplasia and straddling atrioventricular valves were less frequent than reported by Houyel and associates. Ventricular malposition was not present in our initial anatomic series of L-TGA but was seen later[3-5] and was frequent in the other types of the syndrome. Juxtaposition of the atrial appendages was also seen more frequently than in D-TGA.

Absence of the left coronary artery was not a feature of our cases of L-TGA, and in other types of the syndrome there were few cases of single left coronary artery and no cases of single right coronary artery. We noticed, however, that absence of the coronary sinus without splenic anomalies was surprisingly common in our cases.

The surgical implications of these anatomic peculiarities were analyzed in previous reports,[4, 6] but surgical advances since then (foremost among them the contributions of Dr. Planché's group) may have rendered obsolete some of the previous considerations. However, our surgical experience with older techniques in these patients was very satisfactory, with no hospital deaths (three of four operations on this group of patients[3] were performed by Aldo R. Castañeda at Children's Hospital Medical Center, Boston, Mass.). Lincoln and associates,[6] using similar techniques, also reported survival at least 1 year after the operation in all their patients.

I congratulate the authors on their successful surgical experience and fully agree with them on the statement that L-TGA deserves special attention among the various forms of TGA with atrioventricular concordance.

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Reply to the Editor:

We thank Otero-Coto for his interest in our article,[1] for his comments, and for drawing attention to his own work on hearts with {S,D,L} segmental anatomy.[2-5] Otero-Coto's publications deal with anatomically corrected malposition {S,D,L} (ACM), double-outlet right ventricle {S,D,L} (DORV), transposition of the great arteries {S,D,L} (TGA), and double-outlet left ventricle {S,D,L} (DOLV). He hypothesizes that despite the