Factors associated with increased atrial septum thickness in infants with left heart hypoplasia syndrome: implications for percutaneous septostomy

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Introduction: Hypoplastic Left Heart Syndrome (HLHS) encompasses a spectrum of cardiac malformations characterized by underdevelopment of the left heart structures and aorta that, despite the low incidence among cases of congenital heart disease have high mortality rates. Phenotypic variations of the anomaly may influence the success of the interventional and surgical procedures, especially the characteristics related to the atrial septum and foramen ovale (FO), in cases where balloon catheter septostomy is necessary to maintain the interatrial shunt.

Objectives: To analyze the gross features of the atrial septum of hearts with HLHS in order to define and guide therapeutic procedures on this structure, correlating them with other morphological alterations of the syndrome, such as the anatomy of the heart valves.

Methods: We studied 18 hearts of patients diagnosed and deceased with HLHS, all belonging to the anatomic collection of the Laboratory of Pathology of the Heart Institute (InCor) HCFMUSP. The atrial septum was evaluated for FO patency and size, as well as for the presence of bulging of its lamina to one or the other atrial cavity. Other morphological features of the anatomical specimen such as atresia or patency of the mitral valve and caliber of the ascending aorta and pulmonary trunk were annotated. After resection of the atrial septum, histological sections were made for microscopic measurements of the maximum and minimum thickness of the lamina (atrial septum).

Results: The mean age of the patients was 34.5 days (57% male), with a mean atrial septal thickness of 1.90 mm (0.63-4.09 mm). The mean diameter of the pulmonary trunk and aorta were 1.16 cm and 0.22 cm, respectively. FO was patent in 39% of cases (mean diameter 4,4 mm, range 4 to 8 mm), with bulging of the lamina to one of the atrial cavities in half of the cases. The mitral valve was atretic in 21% of the specimens. There was a significant difference in the mean thickness of the atrial septum in cases with patent versus closed FO, being thicker in cases where the FO was closed (p = 0.047). The ratio FO/age presented a statistically significant negative correlation with the atrial septum thickness (r = -0.76 and p <0.05).

Discussion and Conclusions: The results indicate that the patency and size of the oval foramen may influence the thickness of the atrial septum, suggesting that this factor may limit the success of therapeutic interventions, especially balloon catheter septostomy.

Keywords: Hypoplastic left heart syndrome; Atrial septum; Foramen ovale.