Suggesting that an aggressive treatment should be early proposed.

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Pulmonary arterial hypertension after bone marrow transplantation in children

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Background: Pulmonary arterial hypertension (PAH) is a rare but devastating complication of bone marrow transplantation (BMT). It has been reported after BMT for malignant disease or immune deficiencies.

Methods: Over a period of 10 years, all children with PAH after BMT were reviewed for underlying disease, pre-BMT conditioning, associated hepatic veno-occlusive disease, characteristics of PAH, management of PAH and outcome.

Results: 14 patients were diagnosed to have PAH after BMT (mean age 8.5 months, mean age at BMT 5.7 months). The underlying disease was an immune deficiency in 11 cases and a malignant disease in 3 cases. Eleven patients received chemoprophylaxis before BMT and conditioning was the same in all cases. Seven patients had a hepatic veno-occlusive disease before the onset of PAH. PAH appeared 40 days after BMT (4-96 days). The presenting symptom was dyspnea in 13 cases and syncope in 1 case. Extensive microbiological investigations were negative in all patients. PAH was diagnosed on echocardiography in all cases and confirmed by right heart catheterization in 7 cases. All patients had normal wedge pressure. Acute vasodilation testing with NO° was positive in all cases and confirmed by right heart catheterization in 7 cases. All patients were paced before 10 years old. Pacing was required for symptomatic bradycardia in 37.5% whereas prophylactic cardiac pacing accounted for 61.6%. The median follow-up was 96 months (from 6 to 384 months). 85.1% experienced no complication and neither dilated cardiomyopathy nor death had occurred at last follow-up. Pacemaker-related complications appeared in 11.6%.

Conclusion: We describe the largest reported experience with isolated and non immune congenital and childhood AVB. Such a block is a nodal damage from unknown origin that may postnatally progress in incomplete forms. Outcomes are not influenced by age at diagnosis. Prognosis is very good with no late-onset dilated cardiomyopathy, a few pacemaker-related complications in the modern technological era and no death at last follow-up.

295

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296

Usefulness of three-dimensional echocardiography for the diagnosis of bicuspid aortic valve: a pediatric study

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Background: The bicuspid aortic valve (BAV) is the most common congenital heart defect. Children with BAV are more likely to have valve dysfunction and to require intervention during childhood. According to the subtype of BAV, the prognosis and the treatment may be different.

Objective: The aim of this study was to assess the accuracy of 3D echocardiography (3DE) in order to diagnose BAV and to depict accurately the leaflets morphology.

Methods: 40 consecutive children with suspicion of BAV were included in a prospective monocentric study. 2DE and 3DE views were recorded by the same investigator. Images were reviewed separately by two confirmed pediatric cardiologists in order to assess BAV. Then, we compared 2DE and 3DE for the description of the spatial position of cusps and raphes. The association with aortic aneurysms, aortic valve insufficiency or stenosis and other cardiac malformations were also reported.

Results: The median age was 4.8 years (± 7.3). For 19 patients (47%), 3DE allows a better visualization of the leaflets morphology leading to a reclassification of the BAV. Raphes are better distinguished by 3D live (sensitivity = 100% [CI95%, 88-100], specificity = 100% [CI95%, 87-100]) or biplane echocardiography (sensitivity = 96% [CI95%, 80-99], specificity = 63% [CI95%, 35-85]) than 2DE (sensitivity = 86% [CI95%, 62-96], specificity = 28% [14-50]). Inter-observer variability is almost null.

Conclusion: 3D live and biplane echocardiography is a simple, rapid and reliable method for the diagnosis and the accurate description of BAV in children. This technique may be particularly helpful in order to precise the prognosis or to guide the surgeon.

Keywords: Bicuspid aortic valve, Three-dimensional echocardiography.