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0-1-C1240:aortic valve repair versus replacment in laubry and pezzi syndrome

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Introduction :

- *The Laubry and Pezzi syndrome is a rare congenital heart disease associating ventricular septal defect to aortic regurgitation.
- *In our study we propose to analyze the epidemiological, anatomic, clinical and paraclinical features as well as the evolution of this pathology in Algeria.

méthods :

- *This is a retrospective study on 10 patients with a Laubry and Pezzi syndrome operated at our institution over 09 years.

Résultats :

- *The middle age of discovery of the disease is twenty years with extreme ranging from 6 to 43 years.
- *There was a male predominance (65% boys). Almost all patients are symptomatic.
- *Echocardiography is the essential examination to establish the diagnosis.
- *It was perimembranous in 80% of cases associated with prolapse and leaks aortic degrees.
- *The average age at surgery was 20 years. VSD was closed in 100% of patients and associated with conservative valve repair in 6 other.
- *Four patients underwent aortic valve replacement using mechanical prosthesis. The immediate evolution was good in the majority of cases.
- *The mean long-term is 3 years.
- *For the patients who underwent closure of VSD without valvular gesture, 03 have saw their AR disappear, 02 have increased their AR become moderate requiring further surgery on the aortic valve and the remaining 01 retained their grade 1 AR in the long-term follow-up.

Conclusion :

- *The diagnosis and the regular and frequent monitoring of high VSD including infundibular and périmembranous one must be systematic.
- *In fact, the onset of AR during the evolution of a VSD is a turning progression of the disease since its occurrence ecompromises the long-term prognosis and modifies the therapeutic indications.

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