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Results of delayed surgical treatment in children with tetralogy of Fallot from a devloping country

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Introduction: Corrective surgery of tetralogy of Fallot (TOF) is usually performed in the first year of life. Treatment options in developing countries are frequently restricted due to limited availability of diagnostic and therapeutic facilities. This results in a significant number of children and adolescents with native TOF and severe obstruction of the RVOT. We report the results in a cohort of children from Senegal who were referred to our center for surgical treatment.
Patients: From 05/2004 - 10/2016 16 children with TOF were referred with a mean age of 7.0 years (range 0.9-14.8 years). The primary diagnosis established in Senegal by echocardiography was confirmed in all patients. Cardiac catheterization was required in 3/16 patients. Almost all patients were dystrophic (mean BMI 13.7, range 9.8 – 16.7). All patients were significantly cyanosed and presented in NYHA class III-IV.
Results: Primary corrective surgery was performed in 13/16 patients. A valve sparing transatrial approach was possible in 10 patients, one of these patients required secondary transanular patch enlargement 2 days later. One patient, who presented with active endocarditis and vegetations on the aortic and pulmonary valve, underwent corrective surgery including aortic and pulmonary valve replacement. Corrective surgery with RVOT patch-enlargement was performed in further 2. A primary shunt procedure was required due to persistent hypoxemic spells necessitating treatment with noradrenalin in 2 (13.8 and 3.3 years) and excessive hypertrophy of both ventricles in 1 case (6.8 years). Two of these patients underwent secondary corrective surgery requiring RVOT patch enlargement in one, the third patient awaits corrective surgery. All patients were discharged in good condition (median 12 postoperative days).
Conclusion: Despite of late referral and severe obstruction of the RVOT corrective surgery was possible with low mortality. Preservation of the pulmonary valve was possible despite of severe obstruction of the RVOT in the majority of our patients avoiding the long term sequelae of significant pulmonary regurgitation. Palliative procedures remained reserved for patients with persistent hypoxemic spells and excessive RV hypertrophy. The unfavorable natural history and the disabling sequelae of chronic hypoxemia justify all efforts to attempt late corrective surgery in children with TOF.