TETRALOGY OF FALLOT. PARTICULARITIES IN EVOLUTION ACCORDING TO THE DEGREE OF SEVERITY OF PULMONARY STENOSIS

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Background. Tetralogy of Fallot (TOF) is the most common cyanotic congenital heart defect, representing about 10% of all congenital heart defects which comprises four structural defects: ventricular septal defect, pulmonary stenosis, overriding aorta, right ventricular hypertrophy. The degree of severity of pulmonary stenosis hugely affects the patient's presentation, management and evolution. **Objective of the study.** To analyze the particularities in evolution of TOF according to the degree of severity of pulmonary stenosis and its significance on management and long-term follow ups. **Material and methods.** Literature reviews on TOF according to mild, moderate and severe degree of pulmonary stenosis were bought into study and its management measures based on clinical data, paraclinical investigations and outcomes were recorded. The evolution, course, and prognosis of the classified groups was determined through analysis. **Results.** Evaluation of pulmonary stenosis using echocardiography had been done for increased visualization of the pulmonic valve. Most patients with mild pulmonary stenosis are asymptomatic, moderate or severe patients who experience dyspnea on exertion or associated fatigue, depending on the severity of the obstruction. Infants with critical pulmonary stenosis presented with cyanosis after birth. Studies show that patients with mild or moderate stenosis had better outcomes and less mortality than severe stenosis which have poor outcomes and higher mortality. The less severe stenosis is well tolerated than severe stenosis in neonatal ductal dependent stenosis. **Conclusion.** Catheter-based therapy is corrective for the majority of patients with isolated valvular pulmonary stenosis and surgical therapy in patients with subvalvular and supravalvular obstruction and use of a catheter-based intervention during continued follow-up and surveillance of these patients. **Keywords:** tetralogy of Fallot, pulmonary stenosis, evolution, outcomes, prognosis.

TREATMENT STRATEGY FOR SEVERE COMBINED IMMUNODEFICIENCY IN CHILDREN

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Background. Severe Combined Immunodeficiency (SCID) is a rare but severe pediatric disease that is marked by a severe deficiency of T and B lymphocytes, which results in a high susceptibility to infections. **Objective of the study.** The purpose of this research is to assess current developments in management of SCID in children by analyzing key trends, clinical findings, and difficulties from academic publications published between 2019 and 2024. **Material and methods.** A literature review with a primary focus on clinical trials, meta-analyses, and peer-reviewed articles from 2019 to 2024 was conducted. The search terms "Severe Combined Immunodeficiency", "SCID", "pediatric immunodeficiency," "gene therapy," and "hematopoietic stem cell transplantation" were combined to search PubMed, MEDLINE, and the Cochrane Library. **Results.** Increased use of newborn screening allows for early diagnosis in these patients. Protective isolation, precautions related to transfusion of blood products, avoidance of live vaccines, and continuous prophylactic anti-infectious treatment should be included in initial management before a definitive diagnosis is made. The gold standard for curative care is still hematopoietic stem cell transplantation (HSCT), with better results now possible because of developments in donor matching and conditioning regimens. Gene therapy as an alternative has emerged, offering the potential for curative therapy for specific genetic variants of SCID. Studies indicate that gene therapy has a high success rate in correcting immune deficiencies. **Conclusion.** Newborn screening, HSCT advancements, and gene therapy have made significant advances in understanding and treating SCID in the previous five years. However, chronic illnesses need continuing research and international cooperation owing to treatment availability and long-term management difficulties. **Keywords:** SCID, gene therapy, hematopoietic stem cell transplantation