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CLINICAL AND INSTRUMENTAL CHARACTERISTICS OF THE COURSE AND PROGNOSIS IN CHILDREN WITH TETRALOGY OF FALLOT

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ABSTRACT: English: Background: Tetralogy of Fallot (ToF) is the most common cyanotic congenital heart defect (CHD). The clinical course and prognosis depend heavily on the degree of right ventricular outflow tract obstruction (RVOTO). Objective: To study the clinical and instrumental features of Tetralogy of Fallot in children in the Andijan region and to determine prognostic factors affecting the disease course. Methods: A retrospective analysis of 60 children aged 6 months to 5 years diagnosed with ToF was conducted. Patients were assessed using pulse oximetry, ECG, and Doppler Echocardiography. The McGoon ratio was used to assess pulmonary artery anatomy. Results: The study identified that 75% of patients presented with "classic" cyanotic forms, while 25% had "pale" forms. A strong inverse correlation was found between the McGoon ratio and the frequency of hypoxic spells (r = -0.72). Children with a McGoon ratio <1.2 had a significantly worse prognosis and higher pre-operative morbidity. Conclusion: The clinical severity of ToF is directly related to the morphometric parameters of the pulmonary arteries. Early instrumental assessment allows for risk stratification and timely surgical intervention, improving the long-term prognosis.
Keywords: Tetralogy of Fallot, congenital heart defects, echocardiography, hypoxic spells, prognosis, pediatrics.

FALLO TETRADASI BO'LGAN BOLALARDA KASALLIK KECHISHI VA PROGNOZINING KLINIK-INSTRUMENTAL TAVSIFI

ANNOTATSIYA: Kirish: Fallo tetradasi (FT) ko'k o'tkir tug'ma yurak nuqsonlari (TYN) ichida eng ko'p uchraydigan turidir. Kasallikning klinik kechishi va prognozi o'ng qorincha chiqish qismi obstruksiyasi (O'QChQO) darajasiga bog'liq. Maqsad: Andijon viloyatidagi bolalarda Fallo tetradasining klinik va instrumental xususiyatlarini o'rganish hamda kasallik kechishiga ta'sir etuvchi prognostik omillarni aniqlash. Usullar: FT tashxisi qo'yilgan 6 oylikdan 5 yoshgacha bo'lgan 60 nafar bolaning retrospektiv tahlili o'tkazildi. Bemorlar pulsoksimetriya, EKG va Doppler Exokardiografiya yordamida tekshirildi. O'pka arteriyasi anatomiyasini baholash uchun McGoon indeksi qo'llanildi. Natijalar: Tadqiqot shuni ko'rsatdiki, bemorlarning

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75 foizida "klassik" ko‘kargan shakllar, 25 foizida esa "oq" shakllar kuzatildi. McGoon indeksi va gipoksik xurujlar chastotasi o‘rtasida kuchli teskari korrelyatsiya aniqlandi (r = -0,72). McGoon indeksi <1,2 bo‘lgan bolalarda prognoz sezilarli darajada yomonroq va operatsiyadan oldingi kasallanish darajasi yuqori bo‘ldi. Xulosa: FTning klinik og‘irligi o‘pka arteriyalarining morfometrik ko‘rsatkichlari bilan bevosita bog‘liq. Erta instrumental baholash xavf guruhlarini ajratish va jarrohlik amaliyotini o‘z vaqtida o‘tkazish imkonini beradi, bu esa uzoq muddatli prognozni yaxshilaydi.
Kalit so‘zlar: Fallo tetradasi, tug‘ma yurak nuqsonlari, exokardiografiya, gipoksik xurujlar, prognoz, pediatriya.

КЛИНИКО-ИНСТРУМЕНТАЛЬНАЯ ХАРАКТЕРИСТИКА ТЕЧЕНИЯ И ПРОГНОЗ У ДЕТЕЙ С ТЕТРАДОЙ ФАЛЛО

АННОТАЦИЯ: Введение: Тетрада Фалло (ТФ) является наиболее распространенным синим врожденным пороком сердца (ВПС). Клиническое течение и прогноз в значительной степени зависят от степени обструкции выходного тракта правого желудочка (ВОПЖ). Цель: Изучить клинико-инструментальные особенности Тетрады Фалло у детей Андижанской области и определить прогностические факторы, влияющие на течение заболевания. Методы: Проведен ретроспективный анализ 60 детей в возрасте от 6 месяцев до 5 лет с диагнозом ТФ. Пациенты оценивались с помощью пульсоксиметрии, ЭКГ и допплер-эхокардиографии. Для оценки анатомии легочной артерии использовался индекс МакГуна. Результаты: Исследование выявило, что у 75% пациентов наблюдались «классические» цианотичные формы, а у 25% — «бледные» формы. Обнаружена сильная обратная корреляция между индексом МакГуна и частотой одышечно-цианотических приступов (r = -0,72). Дети с индексом МакГуна <1,2 имели значительно худший прогноз и более высокую предоперационную заболеваемость. Заключение: Клиническая тяжесть ТФ напрямую связана с морфометрическими параметрами легочных артерий. Ранняя инструментальная оценка позволяет стратифицировать риски и своевременно проводить хирургическое вмешательство, улучшая отдаленный прогноз.
Ключевые слова: Тетрада Фалло, врожденные пороки сердца, эхокардиография, одышечно-цианотические приступы, прогноз, педиатрия.

INTRODUCTION

Tetralogy of Fallot (ToF) is one of the most prevalent and clinically significant congenital heart defects (CHD), accounting for approximately 7-10% of all congenital heart anomalies. Anatomically, it is characterized by four classic features: a large ventricular septal defect (VSD), obstruction of the right ventricular outflow tract (RVOTO), overriding of the aorta, and right ventricular hypertrophy.

The natural history and prognosis of ToF are largely dictated by the severity of the pulmonary stenosis. In the absence of surgical intervention, the mortality rate is high, with survival rates dropping significantly after the first year of life. In Uzbekistan, and particularly in the Andijan region, improvements in diagnostic capabilities have allowed for earlier detection, yet challenges

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remain in the timely management of complex cases presenting with severe hypoplasia of the pulmonary arteries.

The clinical presentation varies from mild cyanosis ("pink ToF") to severe hypoxia with life-threatening "tet spells" (hypercyanotic episodes). Accurately characterizing these clinical features and correlating them with instrumental findings (specifically echocardiographic parameters) is crucial for determining the optimal timing for surgery and predicting outcomes. This study aims to provide a comprehensive clinical-instrumental analysis of ToF in the local pediatric population to improve prognostic accuracy.

LITERATURE REVIEW

Pathophysiology and Clinical Variance: Classic literature establishes that the magnitude of the right-to-left shunt in ToF is determined by the ratio of resistance to flow across the RVOTO versus the systemic vascular resistance (SVR). Starr et al. (2019) emphasized that children with severe infundibular stenosis are prone to frequent hypoxic spells, which are major predictors of cerebral complications.

Instrumental Diagnostics: Echocardiography (EchoCG) remains the gold standard for preoperative assessment. Key parameters include the pressure gradient across the pulmonary valve and the anatomy of the pulmonary arteries. The McGoon ratio (diameter of distal right and left pulmonary arteries divided by the diameter of the descending aorta at the diaphragm level) and the Nakata index are critical prognostic markers. Studies by Bokeria et al. (2020) in similar demographic settings suggest that a McGoon ratio of less than 1.2 indicates unfavorable anatomy for primary radical correction, necessitating palliative shunting.

Prognostic Factors: According to Van der Ven et al. (2021), factors such as low birth weight, early onset of cyanosis (<3 months), and significant polycythemia (Hematocrit >60%) are associated with a more complicated postoperative course. However, there is limited data specifically correlating these instrumental indices with the pre-surgical clinical burden in the pediatric population of the Fergana Valley.

MATERIALS AND METHODS

Study Design - A retrospective observational study was conducted at the Department of Hospital Pediatrics, Andijan State Medical Institute, analyzing patient records from 2022 to 2024.

Participants - The study included 60 children (34 boys, 26 girls) aged 6 months to 5 years with a confirmed diagnosis of Tetralogy of Fallot. Inclusion Criteria: Diagnosis confirmed by Doppler EchoCG, absence of other major non-cardiac malformations. Exclusion Criteria: Pulmonary atresia with VSD (as a distinct extreme form), AV canal defects.

Clinical Assessment - Evaluation of cyanosis severity (saturation SpO2 at rest and activity), frequency of hypoxic spells (per week/month), and physical development (height/weight percentiles).

ECG: Assessment of Right Ventricular Hypertrophy (RVH) signs.

Doppler Echocardiography - Performed using standard views. Measured parameters: VSD size, RVOTO pressure gradient (PG), Pulmonary Artery (PA) annulus size, and the McGoon ratio.

Statistical Analysis - Data were analyzed using IBM SPSS Statistics v.26. Pearson correlation coefficients (r) were calculated to assess relationships between instrumental parameters and clinical severity. Differences between groups were analyzed using the Student's t-test.

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RESULTS

Clinical Characteristics: The patients were categorized into two groups based on clinical presentation: Group A (Severe/Cyanotic): SpO2 < 85%, frequent spells (n=45). Group B (Moderate/Pink): SpO2 ≥ 85%, rare or no spells (n=15).

As shown in Table 1, Group A patients had significantly higher hematocrit levels (compensatory polycythemia) and lower physical development indices.

Table 1: Clinical and Laboratory Characteristics of Patients

Parameter	Group A (Severe) (n=45)	Group B (Moderate) (n=15)	P-value
Mean Age (months)	28.4 ± 4.2	32.1 ± 5.1	>0.05
SpO2 at rest (%)	76.5 ± 3.4	88.2 ± 2.1	**<0.001**
Hematocrit (%)	58.4 ± 4.5	44.2 ± 3.1	**<0.001**
Hypoxic Spells (history)	88.9% (40/45)	13.3% (2/15)	**<0.001**
Physical Development (<3rd percentile)	62.2%	20.0%	**<0.01**

Instrumental Findings and Prognosis: Echocardiographic analysis revealed distinct anatomical differences. The pressure gradient across the RVOT was paradoxically lower in some very severe cases due to decreased flow, but the anatomical obstruction was severe. The McGoon ratio proved to be the most sensitive marker.

Table 2: Echocardiographic Parameters and Anatomical Prognosis

EchoCG Parameter	Group A (Severe)	Group B (Moderate)	Correlation with Spells (r)
VSD Size (mm)	14.2 ± 2.1	11.5 ± 1.8	0.15 (weak)
Aortic Override (%)	55 ± 5%	40 ± 4%	0.32 (moderate)
Pulmonary Valve Annulus (Z-score)	-3.5 ± 0.5	-1.8 ± 0.4	**-0.68 (strong)**
McGoon Ratio	**1.1 ± 0.2**	**1.8 ± 0.3**	**-0.72 (strong)**

Children in Group A with a McGoon ratio <1.2 (indicating hypoplastic pulmonary arteries) had a poorer prognosis. 80% of children in this category required palliative surgery (Blalock-Taussig shunt) before radical correction could be attempted, whereas children in Group B were candidates for primary repair.

DISCUSSION

The results of this study highlight the critical link between anatomical metrics and clinical outcomes in children with ToF in the Andijan region.

We observed that clinical cyanosis and physical growth retardation are late indicators of disease severity. The fact that 62.2% of children in the severe group were below the 3rd percentile for physical development indicates chronic tissue hypoxia, which could have been mitigated by earlier intervention.

The strong correlation (r = -0.72) between the McGoon ratio and the frequency of hypoxic spells confirms the hypothesis that the development of the pulmonary vascular bed is the primary

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determinant of prognosis. Our findings align with global standards suggesting that a McGoon ratio <1.2 is a "red flag" for poor prognosis. In our setting, these patients represent the most challenging cohort, requiring staged surgical approaches.
Furthermore, the high hematocrit levels in Group A serve as a double-edged sword: while compensating for hypoxia, they increase blood viscosity, raising the risk of thrombotic complications, which significantly worsens the prognosis.

CONCLUSION

Based on the clinical-instrumental analysis of 60 pediatric patients with Tetralogy of Fallot, we conclude:
ToF presents with a wide spectrum of severity. The presence of frequent hypoxic spells and polycythemia (Hct >55%) are absolute indicators of severe hemodynamics requiring urgent evaluation.
The McGoon ratio is a highly reliable prognostic tool. A value below 1.2 is strongly associated with severe cyanosis, growth failure, and the need for palliative rather than radical initial surgery.
Chronic hypoxia in uncorrected ToF leads to significant physical developmental delay in over 60% of severe cases in our region.

RECOMMENDATIONS

Doppler Echocardiography with mandatory calculation of the McGoon ratio and Pulmonary Valve Z-scores should be standard protocol for all children suspected of ToF in Andijan.
Children with "Pink ToF" must be monitored closely, as their relatively mild symptoms may mask significant anatomical defects that worsen over time.

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