

Clinical Significance of Thymomegaly in Children with Congenital Heart Defects

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Abstract

Children with thymomegaly exhibit impaired adaptive capacity associated with functional changes in the hypothalamic–pituitary–adrenal system, while their immunological insufficiency contributes to the development of infectious complications. They represent a group with reduced biological system reliability; therefore, assessment of thymic status is essential for improving outcomes of surgical correction of congenital heart defects in children.

Key words:

Background

Congenital heart defects and great vessel anomalies occur in 0.8–1.2% of all newborns and rank first among causes of mortality due to congenital malformations. In CHD, severe anatomical abnormalities of the heart and great vessels are combined with pronounced hemodynamic disturbances and chronic tissue hypoxia, including organs of the lymphoid system [1,2]. Therefore, a significant proportion of postoperative complications following cardiac surgery is associated with impaired immune system function.

According to the literature, children with CHD often present with enlargement of the thymus gland [3,4]. In most cases, thymomegaly causes technical difficulties during surgical correction of CHD, resulting in the need for partial thymectomy. Treatment strategy and prognosis in patients with CHD largely depend on immunological parameters directly related to the structure and function of the thymus.

The short life expectancy and high mortality rates among young children with CHD emphasize the relevance of this problem and necessitate further in-depth research aimed at improving diagnostic approaches and developing prognostic criteria.

Materials and Methods

A retrospective analysis of medical records of deceased patients with CHD was performed, along with a clinical and instrumental examination of 84 children aged 1 month to 3 years with VSD and Tetralogy of Fallot, including 54 patients with thymomegaly and 30 children without thymic pathology. Diagnosis of CHD was based on medical history, physical examination, and findings from phonocardiography, electrocardiography, and echocardiography. Thymic enlargement was confirmed by radiographic and ultrasound studies. The diagnosis of thymomegaly was established based on the cardiomyocardial-thoracic index (CTTI) and vasocardial index (VCI). The thymus was considered enlarged when $CTTI \geq 0.33$ and $VCI \geq 60\%$.

Results and Discussion

To assess the role of thymic pathology in the mortality structure of young children, a retrospective analysis of medical records and autopsy reports of deceased children from 1999 to 2001 at the Republican Scientific Center of Emergency Medical Care (Ministry of Health of the Republic of Uzbekistan) was conducted. The analysis revealed thymic pathology in all cases, with thymomegaly accounting for 53.3%, accidental thymic involution for 26.6%, thymic hypoplasia for 13.3%, and thymic agenesis for 6.6%. In 60% of cases, thymic pathology and CHD were accompanied by multiple congenital malformations of the immune system, spine, and gastrointestinal tract. The combination of CHD and multiple congenital anomalies led to deterioration of the child's condition and death within 24 hours in 63.3% of children with thymomegaly, whereas in children with other thymic pathologies, mortality occurred on days 7–14 of hospitalization in 100% of cases.

To identify clinical features, patients with CHD associated with thymomegaly were compared with those without thymomegaly. Among children with CHD and thymomegaly, infants under one year of age predominated (44.4%), whereas among children without thymomegaly, children in the third year of life prevailed (53.3%).

Analysis of maternal obstetric history and anamnestic data of children with thymomegaly showed a higher incidence of gestosis throughout pregnancy ($p < 0.05$), perinatal central nervous system damage ($p < 0.05$), grade I–II anemia ($p < 0.05$), nutritional disorders (low body weight), and a high prevalence of allergic burden ($p < 0.05$) compared to children without thymic pathology. In the group without thymomegaly, delayed psychomotor development and gestosis during the first half of pregnancy were more frequent ($p < 0.05$).

Congenital anomalies of other organs were detected in 46.3% of children with thymomegaly, including inguinoscrotal hernia (3.7%), umbilical hernia (1.9%), cleft palate (1.9%), syndactyly of toes (1.9%), high-arched palate (13%), malformed auricles (22.2%), and Down syndrome in one patient (1.9%).

In the preoperative period, children with VSD and thymomegaly demonstrated a high morbidity rate. By the time of surgical correction, 66.7% of patients had frequent recurrent respiratory infections (acute respiratory viral infections, bronchitis, pneumonia), classifying them as frequently ill children. In contrast, this rate was 26.7% among children with VSD without thymomegaly. Despite young age, pulmonary hypertension grade II–III was observed in 42.9% of children with VSD and thymomegaly, indicating early development of this complication and the need for urgent surgical intervention. No cases of severe pulmonary hypertension were observed in children without thymomegaly.

In children with Tetralogy of Fallot, disease severity was associated with dyspneic-cyanotic spells. In patients with thymomegaly, these episodes occurred 2–3 times daily in 23.1% of cases, daily in 53.8%, and 2–3 times per month in the remaining cases. In children without thymomegaly, attacks occurred only 1–2 times per month.

In the postoperative period, the general condition of children in both groups corresponded to the severity of surgery. However, the dynamics of clinical recovery differed significantly: improvement was observed on average by postoperative day 5 in children without thymomegaly, whereas in children with thymomegaly improvement occurred only by day 10.

Postoperative complications of hemodynamic and infectious nature were observed in 38.8% of children with CHD and thymomegaly, including postoperative pneumonia (14.8%), post-cardiotomy syndrome (13%), surgical wound infection (7.4%), acute heart failure (3.7%), rhythm disturbances (atrioventricular block) (3.7%), and disseminated intravascular coagulation syndrome (1.9%). In children without thymomegaly, only two cases (6.7%) of post-cardiotomy syndrome were registered; the postoperative course in the remaining patients was relatively uncomplicated, and no deaths were recorded.

The mortality rate in the group with thymomegaly was 9.3%, including one patient with VSD and four with Tetralogy of Fallot. The main causes of death were acute heart failure (3.7%), multiple organ failure (1.9%), disseminated intravascular coagulation syndrome (1.9%), and cardiac rhythm disturbances associated with bronchopneumonia (3.7%).

These findings confirm that children with thymomegaly have impaired adaptive capacity associated with functional alterations of the hypothalamic–pituitary–adrenal system, and that their immunological insufficiency contributes to the development of infectious complications.

Conclusions

1. Retrospective analysis of medical records of deceased patients with CHD revealed various thymic pathologies, with thymomegaly predominating (53.3%).
2. Children with thymic enlargement belong to a group with low biological system reliability; therefore, thymic status should be considered to improve surgical outcomes in children with CHD.
3. Ultrasonographic and radiographic assessment of the thymus is recommended for all children with CHD to detect thymomegaly.

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