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Infective endocarditis in a boy with Down syndrome after cardiac surgery in infancy and removal of 13 teeth in the fifth year of life

Infekcyjne zapalenie wsierdzia u chłopca z zespołem Downa po operacji kardiochirurgicznej w okresie niemowlęcym i usunięciu 13 zębów w piątym roku życia

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Abstract

A 14-year-old boy with Down's syndrome and an atrioventricular septum defect underwent cardiosurgical treatment at 3 months of age. The postoperative course included pericardiocentesis due to fluid accumulation, anemia, lack of body weight increase, and kidney failure. In the years that followed, the boy's condition was stable, although he often suffered from infections. The reconstructed mitral valve showed good function, the leaflets were not thickened. At 5 years of age, the patient underwent extraction of 13 teeth consecutively, due to extensive inflammatory changes, without antibiotic cover. The child's health deteriorated after the procedure. Inflammation indicators were positive and symptoms of sepsis appeared. Physical examination revealed splenomegaly, and *Streptococcus mitis* and *Staphylococcus hominis* bacteria were cultured in the blood culture. Transthoracic echocardiography and transesophageal echocardiography showed a hyperechogenic structure of 10 × 4 mm on the mitral valve, causing a narrowing of the inflow to the left ventricle and an exacerbation of grade I to III valve regurgitation as well as features of pulmonary hypertension. Vegetation in the course of infectious endocarditis was suspected and intensive antibiotic therapy was initiated in accordance with the antibiogram. After obtaining sterile cultures, the boy underwent diagnostic cardiac catheterization, which showed post-capillary type reversible pulmonary hypertension. The child was qualified for cardiac surgery during which the vegetation was removed and the Medtronic Hancock 23 mm biological valve was implanted in the mitral field. The boy's condition has systematically improved and he is currently under outpatient control. After several years of follow-up, the mitral valve shows slight regurgitation. Also, a mild obstruction of the left ventricle outflow tract is observed, not requiring any intervention at the moment.

Key words: Down's syndrome, caries dentum, infective endocarditis, vegetation, biological heart valve

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Introduction

Down's syndrome (DS) is present in nearly 10% of all children undergoing cardiac surgery, including approximately 2% of neonatal cases [1], while approximately 50% of newborns with DS have congenital heart disease [2]. The most common defect is atrioventricular septum defect, which should be treated surgically before 6 months of age due to the risk of pulmonary hypertension (PH). It is known that children with DS are also more likely to develop infections due to abnormalities of the immune system with T and B cell lymphopenia [3]. The outcome of cardiac surgery is similar to those in the population of children without DS however a greater risk of complications in the group of patients with DS is mainly associated with recurrent infections and comorbidities [4]. Recent studies indicate differences in the oral microbiome in children with DS, which may contribute to an increased incidence of periodontitis [5, 6]. The most common qualification criteria for tooth extractions in DS patients are dental caries, pulpal diseases, and periodontal infections. Due to the higher risk of infective endocarditis (IE), most often caused by a streptococcal infection during the extraction of diseased teeth, such patients should undergo prophylactic antibiotics supplementation [7, 8].

Case report

A 14-year-old boy with DS underwent surgical atrioventricular septum defect correction using the two-patch technique at 3 months of age. The postoperative course included pericardiocentesis due to fluid accumulation, anemia, and renal failure treated in the nephrology ward.

In the years that followed, the boy's condition was stable, although he often suffered from infections. The reconstructed mitral valve showed good function, the leaflets were not thickened (Figure 1). At 5 years of age, the patient developed extensive purulent inflammatory changes in dentition, requiring extraction. At the mother's request, the dentist removed as many as 13 teeth under general anesthesia, but without applying prophylactic antibiotics. After the extraction, the boy required hospital treatment as he developed sepsis symptoms with elevated inflammation indicators, fever, chills, and an enlarged spleen. *Streptococcus mitis* and *Staphylococcus hominis* were cultured from the blood. Transthoracic and transesophageal echocardiographic examinations revealed vegetation of approximately 10 × 4 mm on the anterior leaflet of the reconstructed mitral valve, causing its narrowing with an acceleration of the inflow to the left ventricle and significant III degree regurgitation with features of PH (Figure 2). Intensive antibiotic treatment with amoxicillin was initiated in accordance with the antibiogram. After obtaining negative blood cultures, the patient was qualified to undergo

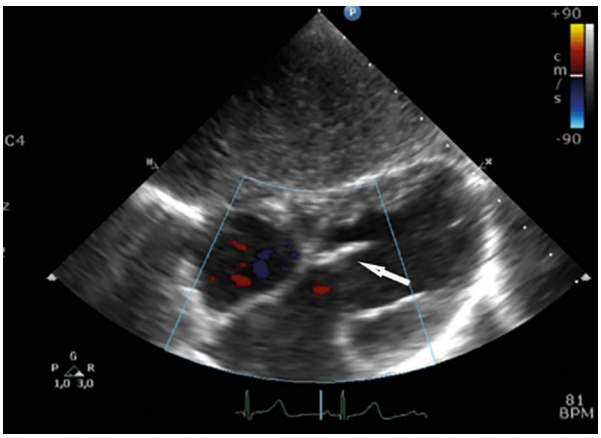

Figure 1. A 3-year-old boy after cardiosurgical treatment of an atrioventricular septal defect. Echocardiographic examination in the 3-chamber sub-sternal view. Mitral valve leaflets without vegetation (arrow)

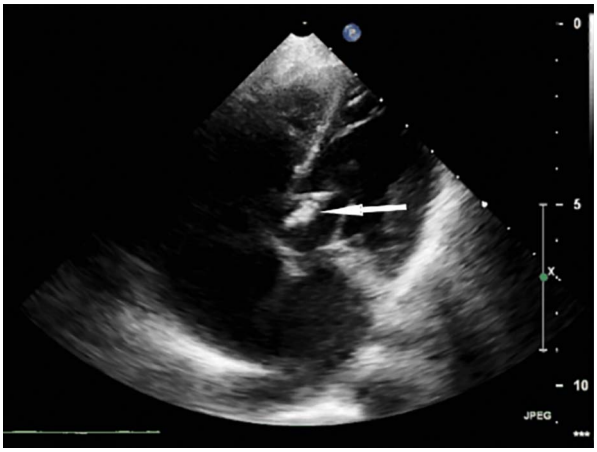

Figure 2. The same patient, 5 years old, after extraction of 13 teeth. Echocardiographic examination in the 5-chamber apex view. Vegetation in the area of the anterior mitral valve leaflet 10 × 4 mm (arrow)

a diagnostic catheterization of the heart, which showed that PH was postcapillary type and reversible. The boy was qualified for cardiac surgery, during which the vegetation was removed and the Medtronic Hancock 23 mm biological valve was implanted in the mitral field. Immediately after the operation, the child developed atrial flutter, effectively treated with cardioversion. He was also treated with antibiotics (vancomycin, imipenem), cardiac drugs (dopamine, corotrope, captopril), diuretics (furosemide), and oral anticoagulant (warfarin). After 6 weeks he was discharged home in good condition. Since then he has been under constant outpatient control regarding both cardiology and dental clinics. Follow-up assesses the function of the implanted mitral valve as satisfactory (Figure 3).

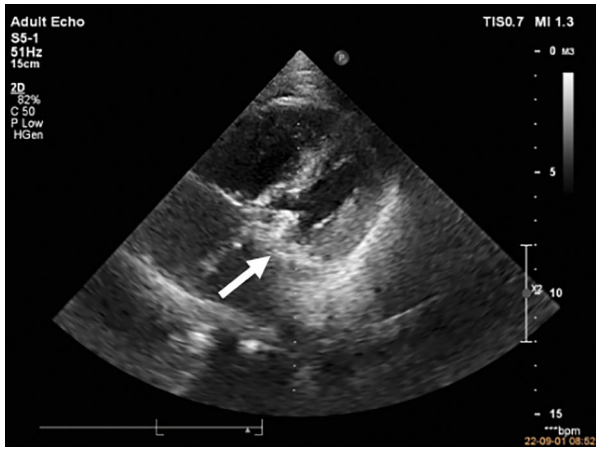

Figure 3. Echocardiographic image in 4-chamber view, 9 years after implantation of the Medtronic Hancock 23 mm biological valve in the mitral position (arrow). The normal size of the ventricles and atria confirms proper valve function

Discussion

The presented course of the disease of a child with DS is apparently typical, but there are a few issues that need to be discussed. It is known that these children are at risk of infections of the oral mucosa and purulent teeth [6, 7]. According to the recommendations of the European Society of Cardiology, the risk group of IE includes patients with implanted artificial valves or with artificial material used for surgical valve repair. In our patient, the correction of the heart defect was performed using the "two-patch technique", i.e. the interventricular defect was closed with a plastic patch, and the interatrial septum was reconstructed with a patch from the pericardium. Thus, according to the European Society of Cardiology guidelines, the use of an artificial patch qualified the boy for the high-risk group of IE [8]. Dental recommendations regarding the current indications for antibiotic prophylaxis in patients after correction of congenital heart defects are based on the standards of IE prophylaxis developed by experts in 2015 and included in the guidelines of the European Society of Cardiology (8). However, more and more attention is paid to the deviation from the prophylactic use of antibiotics in favor of increasing oral hygiene. Especially when it concerns children with congenital heart defects and after implanting artificial materials [9]. It is also known that DS patients are almost panicky about dental checkups and the lack of cooperation in this area is well known. According to the work of Grisolia et al. [10] the level of fear towards the dentist (dental anxiety-DA) is the greater the younger the child is. The prevalence of DA in preschool children is 36.5%, 25.8% in school and 13.3% in school age. It was also noted that, as shown by the meta-analysis, DA is a common problem in children aged 3–18 all over the world [10]. In addition, there are no specialist centers treating teeth in such patients, as the procedures almost always require general anesthesia. As a result, children with DS face great difficulties in dental care. In the described case, the dentist enlisted the help of an anesthesiologist and, with the mother's consent, removed a significant number of sick teeth from the boy, which could be a source of IE, vegetation and valve damage. Today we know that the dentist's decision to remove a large number of teeth at one time was not the best, especially in the case of IE prophylaxis. What's more the lack of antibiotic prophylaxis on purulent tooth infection in a child with a high risk of IE should be considered a mistake, disregarding the fact that the mother herself demanded such a procedure. The boy still remains in the high-risk group of IE, as a biological valve was implanted in place of the mitral valve damaged by vegetation. The available literature does not provide guidelines on how many teeth can be removed in a child with DS at one time, and after cardiac surgery. However, there are guidelines for the use of IE prophylaxis in the group of the highest risk [8] and this situation could have been avoided if standards were to be followed. The replacement of the damaged left-sided valve due to IE for the Medtronic Hancock 23 mm biological valve was undertaken by a team of cardiac surgeons. The choice of this method of treatment turned out to be correct, because after 9 years of observation, satisfactory valve function is found, and the only abnormality is its moderate regurgitation of the valve. At present, there are no indications for reoperation, the heart is fully functional and the boy leads a normal lifestyle. If, however, it was necessary to replace the valve, a modern method of treatment would probably be considered, i.e. hybrid treatment of acute mitral regurgitation in the course of IE with the use of the Melody® valve [11, 12].

Conclusions

Tooth extraction in a child with Down syndrome especially after cardiac surgery carries a high risk of IE, therefore prophylactic antibiotic therapy is recommended.

Conflict of interest

The authors do not report any financial or personal connections with other persons or organizations that could adversely affect the content of the publication and claim the right to this publication.

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Streszczenie

Czternastoletni pacjent z zespołem Downa oraz ubytkiem przegrody przedsionkowo-komorowej był leczony kardiochirurgicznie w 3. miesiącu życia. Przebieg pooperacyjny powikłany był pojawieniem się płynu w osierdziu wymagającego ewakuacji, niedokrwistością, zaburzonym przyrostem masy ciała oraz niewydolnością nerek. W następnych latach życia stan chłopca był stabilny, chociaż często był leczony z powodu infekcji. Zastawka dwudzielna po rekonstrukcji prezentowała dobrą funkcję, płatki nie były pogrubiałe. W 5. roku życia, z powodu rozległych zmian zapalnych uzębienia, pacjent został poddany jednoczasowej ekstrakcji 13 zębów — zabieg wykonano bez osłony antybiotykowej. Po ekstrakcji stan dziecka się pogorszył. Wskaźniki stanu zapalnego były dodatnie, pojawiły się objawy posocznicy. W badaniu fizykalnym stwierdzono splenomegalię, zaś w posiewie krwi wyhodowano bakterie *Streptococcus mitis* oraz *Staphylococcus hominis*. W badaniu echokardiograficznym przezklatkowym i przezprzełykowym, na zastawce mitralnej stwierdzono hiperechogenną strukturę o wymiarach 10 × 4 mm powodującą zwężenie napływu krwi do lewej komory serca, jak również nasilenie niedomykalności zastawki z I na III stopień oraz cechy nadciśnienia płucnego. Podejrzewano wegetację w przebiegu infekcyjnego zapalenie wsierdzia i wdrożono intensywną antybiotykoterapię, zgodną z antybiogramem. Po uzyskaniu jałowych posiewów, u chłopca wykonano diagnostyczne cewnikowanie serca, które wykazało odwracalne pozakapilarne nadciśnienie płucne. Dziecko zakwalifikowano do leczenia kardiochirurgicznego, w czasie którego usunięto wegetację i w pole mitralne wszczepiono zastawkę biologiczną Medtronic Hancock 23 mm. Stan chłopca ulegał systematycznej poprawie i aktualnie pozostaje pod kontrolą ambulatoryjną. W odstępie kilku lat obserwacji zastawka mitralna wykazuje niewielką niedomykalność. Wykazano również zwężenie drogi wypływu z lewej komory, niewymagające obecnie interwencji.

Słowa kluczowe: zespól Downa, uzębienie kariotyczne, infekcyjne zapalenie wsierdzia, wegetacja, zastawka serca biologiczna

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