



# Postpericardiotomy Syndrome in an Infant with Down Syndrome Presenting with Recurrent Pericardial Effusion

Tekrarlayan Perikardial Effüzyon ile Prezente Olan Down Sendromlu Bebeğe Postperikardiyotomi Sendromu

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## Abstract

Postpericardiotomy syndrome (PPS) is a hypersensitivity reaction characterized by inflammation in pleural, pericardial or lung tissue following cardiac surgery or myocardial infarction. Clinically; fever, chest pain, findings related to pleural or pericardial effusion may be seen. In this case report, we wanted to present a case of Down syndrome who was followed up with PPS causing recurrent pericardial effusion after cardiac surgery for 5 months and was successfully treated by opening a pericardial window. PPS is one of the leading causes of post-cardiac morbidity in developed countries, despite all the advances in diagnosis and treatment. The earliest clinical finding is the development of pericardial effusion after the operation. Although pericardiocentesis is performed many times, PPS should be kept in mind in patients with recurrent pericardial effusion, and pericardial window opening should be considered in the treatment.

**Keywords:** Postpericardiotomy syndrome, pericardial effusion, recurrent effusion

## Öz

Postperikardiyotomi sendromu (PPS) kardiyak cerrahi veya miyokard enfarktüsünü takiben plevral, perikardiyal veya akciğer dokusunda enflamasyonla karakterize bir hipersensitivite reaksiyonudur. Klinik olarak; ateş, göğüs ağrısı, plevral veya perikardiyal effüzyona bağlı bulgular görülebilir. Bu olgu sunumunda 5 aylık Down sendromu olan kardiyak cerrahi sonrasında tekrarlayan perikardiyal effüzyona yol açan PPS ile izlenen ve perikardiyal pencere açılarak başarılı bir şekilde tedavi edilen bir olguyu sunmak istedik. PPS, tanı ve tedavi yöntemlerindeki tüm gelişmelere rağmen gelişmiş ülkelerde kardiyak cerrahi sonrası morbiditenin başta gelen nedenlerindedir. En erken klinik bulgusu operasyon sonrası perikardiyal effüzyon gelişimidir. Birçok kez perikardiyosentez işlemi yapılmasına rağmen, tekrarlayan perikardiyal effüzyonu olan hastalarda PPS akılda tutulmalı, perikardiyal pencere açılması tedavide düşünülmelidir.

**Anahtar Kelimeler:** Postperikardiyotomi sendromu, perikardiyal effüzyon, tekrarlayan effüzyon

## Introduction

Postpericardiotomy syndrome is also defined as postacute myocardial syndrome or post cardiac injury syndrome. Postpericardiotomy syndrome is a clinical picture characterized by fever, pleuropericarditis and parenchymal infiltration,

which develops within weeks following pericardial and/ or myocardial injury. This syndrome has been described in the literature after myocardial infarction, cardiac surgery, blunt chest trauma, pacemaker implantation, coronary stent implantation, heart punctures and angioplasty.<sup>1</sup>

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Although the exact etiology of the syndrome is not known, it is accepted that it is an immune event caused by autoantibodies formed against antigens released after pericardial damage.<sup>2</sup>

Pericardial effusion is defined as an increase in fluid between the pericardial leaves. Various factors play a role in the etiology. Tuberculosis (primary cause especially in developing countries), viral infections and postoperative complications may cause effusion.<sup>3</sup>

In this study, we wanted to present a case of postpericardiotomy syndrome, which caused recurrent pericardial effusion resistant to medical treatment after cardiac surgery and required 4 pericardiocentesis and interventional tube insertion, with clinical and radiological findings.

## Case Report

It was learned from the history of the patient, who was diagnosed with complete atrioventricular septal defect for the first time at the age of 4 months, that the patient was born prematurely, had Down syndrome, had body weight below the 3<sup>rd</sup> percentile in the follow-ups, and received thyroid hormone LT4 replacement therapy due to hypothyroidism. Since the atrioventricular valve structure was not suitable for full correction, pulmonary banding surgery was performed 2 weeks after the diagnosis.

A pericardial effusion of 11 mm was detected in the patient who applied to the outpatient clinic with respiratory distress 14 days after discharge. The patient was admitted to the pediatric cardiology service and methylprednisolone, furosemide and angiotensin converting enzyme inhibitor treatments were started.

The patient's blood tests taken at the time of hospitalization were as follows: White blood cell: 5400/mm<sup>3</sup>, polymorphonuclear cell count: 2180/mm<sup>3</sup>, hemoglobin: 11.2 g/dL, C-reactive protein: 4.9 mg/L, albumin: 3.1 g/dL, serum electrolytes: normal. Considering that the etiology of the effusion might be hypothyroidism, the patient's current drug treatment was continued, since the thyroid function test results were between normal intervals, as T4: 1.4 ng/dL and thyroid-stimulating hormone: 4.9 ng/dL. During the follow-up, the patient's pericardial effusion increased and respiratory distress occurred despite the treatment, and the patient's

effusion was drained with an appropriate technique. After the procedure, it was determined with echocardiography that the effusion was not present and the condition of the patient was improved. When it was determined that the pericardial effusion increased to 7 mm on the 4<sup>th</sup> day following the procedure, ibuprofen was started at a dose of 10 mg/kg/day in addition to the steroid. Ten days after the first pericardiocentesis, the patient had tachycardia and tachypnea, and pericardiocentesis was performed for the second time, as a 14.5 mm effusion was observed on echocardiography. When tachycardia (170-180/min) and tachypnea developed during the follow-up of the patient, echocardiography was repeated. Since pericardial fluid did not impair ventricular functions on echocardiography, 55 mL of serous fluid was drained with pericardiocentesis for the third time. Pericardial fluid was in the nature of transudate, as in the first pericardiocentesis (Table 1). Thyroid function tests, metabolic tests, and immunological tests, which were ordered for etiology, were found to be normal. The bone marrow examination was normal in terms of malignancy in the patient, and no malignant cells were seen in the pericardial fluid cytology.

As the patient's respiratory distress continued, he was transferred to the intensive care unit. During the intensive care follow-ups, effusion was drained for the 4<sup>th</sup> time by puncture in the patient who developed tamponade clinic on the 10<sup>th</sup> day. There was no growth in the culture, gram staining was negative (Figures 1a, 1b). After clinically relieved patient re-developed effusion, a pigtail catheter with a 3F intraducer sheath was placed to drain the effusion permanently. Pericardial window was opened because the patient had repeated pericardiocentesis procedures for 4 times in total and effusion did not improve despite the medical treatment given (Figure 2). The patient was discharged asymptotically on the 13<sup>th</sup> day of hospitalization.

## Discussion

Postpericardiotomy syndrome is characterized by fever, chest pain, pericarditis, pleuritis and pneumonia, which usually occur after cardiac trauma.<sup>4</sup> Symptoms are observed in the first 3 weeks after cardiac operations. The two main symptoms of the syndrome are fever and chest pain.<sup>5</sup> Chest

**Table 1. Fluid characteristics after pericardiocentesis**

	Density	Fluid/serum protein	Fluid/serum lactate dehydrogenase	pH	Amount of drained fluid (milliliter)
1. pericardiocentesis	1014	0.4	0.5	7.4	20
2. pericardiocentesis	1015	0.4	0.5	7.5	25
3. pericardiocentesis	1015	0.3	0.4	7.5	55
4. pericardiocentesis	1013	0.4	0.4	7.4	30



**Figure 1a.** Chest X-ray before pericardiocentesis



**Figure 1b.** Chest X-ray after pericardiocentesis

pain in infants can manifest itself as irritability. Almost all cases have pericardial friction rub and most of them have pericardial effusion.<sup>6</sup>

Systemic fluid retention, hepatomegaly, and hypoxemia may be observed. Laboratory examination usually reveals 10-20 thousand leukocytosis and moderately increased sedimentation rate.<sup>7</sup>

Some criteria are used in the diagnosis of postpericardiotomy syndrome. These are classified as major and minor criteria. Major criteria include the presence of pericardial or pleural friction rub, chest pain and fever over 38 degrees. The minor criteria include an increase in erythrocyte sedimentation rate, C-reactive protein level and leukocyte count. It is stated that the presence of two major and one minor criteria is sufficient for diagnosis after excluding the conditions such as pneumonia, heart failure, pulmonary embolism etc.<sup>8</sup>

Although postpericardiotomy syndrome has a self-limiting course in most cases, it carries a potential risk for cardiac tamponade due to the rapid accumulation of pericardial



**Figure 2.** Control chest X-ray after pericardial window operation

fluid. Pericardiocentesis may be required, although it is not common, in the course of the disease. Although late recurrence may be seen, it is very rare. There is almost no data on this syndrome in childhood. In studies conducted to date, the presence of surgical trauma and autoimmune pathogenesis has been blamed as risk factors in the development of the disease.<sup>8</sup> Today, thanks to the developing intensive care units and advanced surgical techniques, the chance of complete surgical correction for congenital heart diseases has increased, which has also increased the incidence of postpericardiotomy syndrome. For this reason, it is very important to know the risk factor or factors for the development of postpericardiotomy syndrome, to prevent complications such as tamponade with close follow-up, early diagnosis and treatment in order to prevent the development of serious complications such as pericardial tamponade in patients with risk factors.<sup>1</sup>

For the reason for the development of pericardial effusion in our patient, it was thought that the patient had postpericardiotomy syndrome, considering the presence of subfebrile fever, irritability, and high acute phase reactants under ibuprofen in the first 3 weeks after the pulmonary banding operation. In addition, the presence of underlying hypothyroidism due to Down syndrome facilitated the development of pericardial effusion.

This syndrome should be considered in patients with recurrent pericardial effusion and congenital heart surgery, and rapid intervention is required to shorten the length of stay in the pediatric intensive care unit.

#### **Ethics**

**Informed Consent:** Verbal consent was obtained from the patient's family.

**Peer-review:** Externally peer-reviewed.

### Authorship Contributions

Concept: Ş.K., M.U.Y., Design: Ş.K., U.A.Ö., Analysis or Interpretation: Ş.K, M.U.Y., U.A.Ö., M.T., Literature Search: Ş.K., M.U.Y., Writing: Ş.K.

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