# Anesthesia Application in A Proteus Syndrome Patient With Pericardial Effusion

## Perikardiyal Efüzyonu Olan Proteus Sendromlu Bir Hastada Anestezi Uygulaması

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## ABSTRACT

Proteus sendromu (PS), Cohen ve Hayden tarafından tanımlanan ve vücutta asimetrik ve aşırı doku büyümesi ile karakterize nadir görülen konjenital bir hastalıktır (1). Bu olgularda ilerleyici iskelet deformiteleri, tümörler, vasküler malformasyonlar, büllöz akciğer hastalığı ve deri lezyonları gibi farklı patolojiler görülebilir. Kardiyak tutulumu inceleyen sınırlı sayıda çalışma vardır.

Bu çalışmada, 18 yaşında PS'lu; tekrarlayan perikardiyal efüzyonu olan erkek hastanın sağ çift lümenli tüp (SÇLT) kullanılarak mini torakotomi ile perikardiyal pencereden başarılı drenajı sunuldu.

**Keywords:** Proteus sendromu, Perikardiyal efüzyon, Sağ çift lümenli tüp

## ÖZ

Proteus syndrome (PS) is a rare congenital disease described by Cohen and Hayden, which is characterized by asymmetric and excess tissue growth in the body (1). Various pathologies such as progressive skeletal deformities, tumors, vascular malformations, bullous pulmonary disease and skin lesions can be seen in these cases. There is a limited number of studies examining cardiac involvement.

In this study, we present an 18-year-old male patient with PS with recurrent pericardial effusion that required mini-thoracotomy using a right double-lumen tube (RDLT) and resulted in successful drainage with a smooth pericardial window.

**Anahtar Sözcükler:** Proteus syndrome, Pericardial effusion, Right double-lumen tube

## INTRODUCTION

PS is a congenital hamartamatous disease frequently affecting the skeletal system, skin, central nervous system (CNS), vascular structures, and soft tissues. PS symptoms occur in childhood and are progressive (1). An informed consent form was obtained from the patient.

## CASE REPORT

An 18-year-old male case was having exertional dyspnea, chest tightness, and shortness of breath. He stated that his complaints had continued for 3 years intermittently and had exacerbated within the previous month. Chest X-ray revealed increased cardio-thoracic ratio (Figure 1), transthoracic echocardiography showed pericardial fluid at the posterior wall at the lateral, at the right atrium, at the right ventricle, and at the apex (Figure 2).

The patient had a history of at least 3 pericardiocentesis procedures due to pericardial effusion. It was decided to open a pericardial window in this case.

On physical examination, respiratory sounds were decreased in the basal part of both lungs. Heart sounds were rhythmically diminished and deep. In terms of face and mouth structure, the nostrils were asymmetrical and deviated slightly to the right, and the placement of the jaw

and teeth was normal. Mallampati and Cormack Lehane scores were II, and neck extension was slightly restricted. There was a less developed shoulder wall compared to the left and color changes that were compatible with cafe–au–lait spots, hand and finger deformities on the same side, plantar skin folding, lymphedema, and deformity in the right foot (Figure 3,4).

General anesthesia was planned, and informed consent was obtained from the case. When the patient was admitted to the operating room, electrocardiogram, noninvasive blood pressure and pulse oximetry monitoring, and preoxygenation with 80% oxygen were applied. The patient was induced with 0.1 mg/kg midazolam, 50 mcg/kg fentanyl, and 0.6 mg/kg rocuronium, and intubated with a 35 F RDLT. The RDLT position was confirmed by auscultation and flexible bronchoscopy. Invasive artery monitorization was performed from the right radial artery, and a central venous catheter was applied from the right internal jugular vein under ultrasound guidance. Anesthesia was maintained with remifentanil, sevoflurane, and rocuronium.

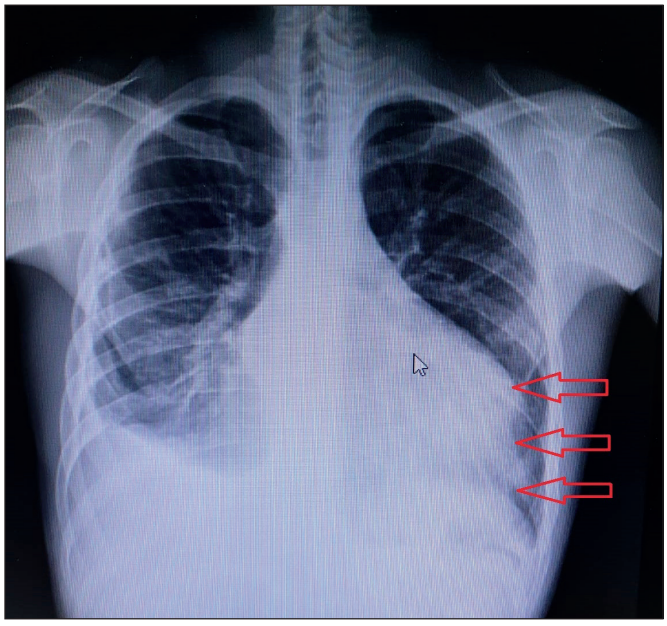

**Figure 1:** PA Chest X-ray image. (⇨: Pleural effusion).

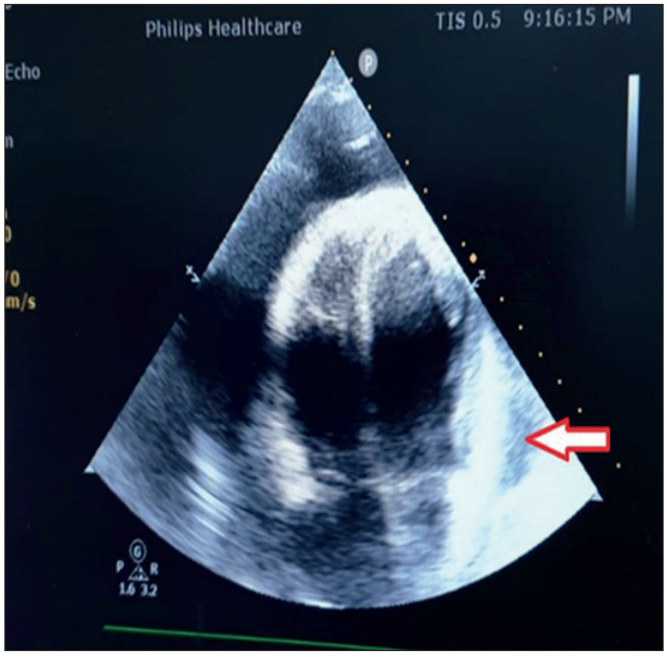


**Figure 2:** Preoperative transthoracic echocardiography image. (⇨: Pleural effusion).

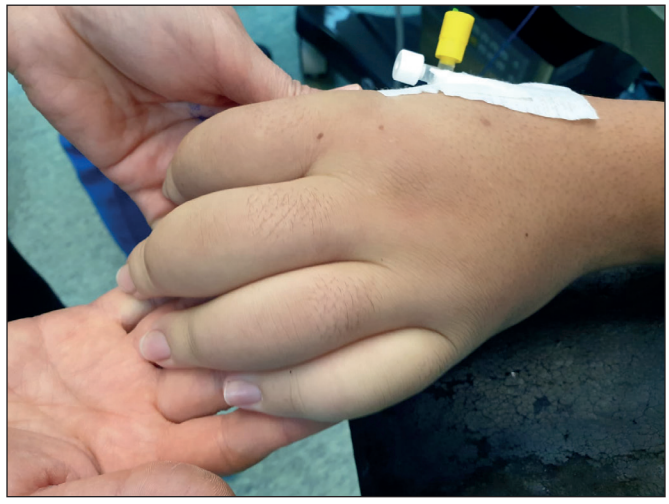

**Figure 3:** Right hand soft tissue hypertrophy.

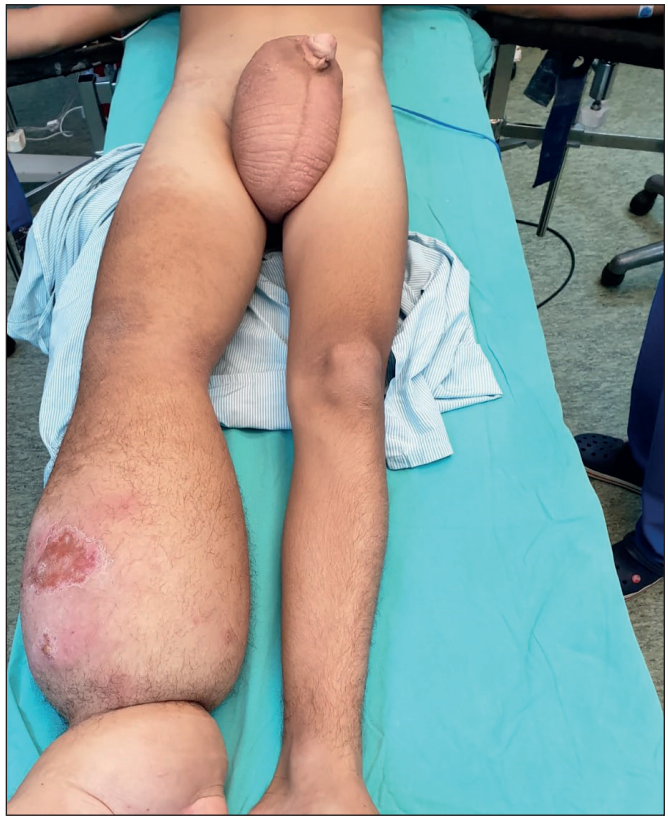

**Figure 4:** Disproportionate excessive growth in the right extremity.

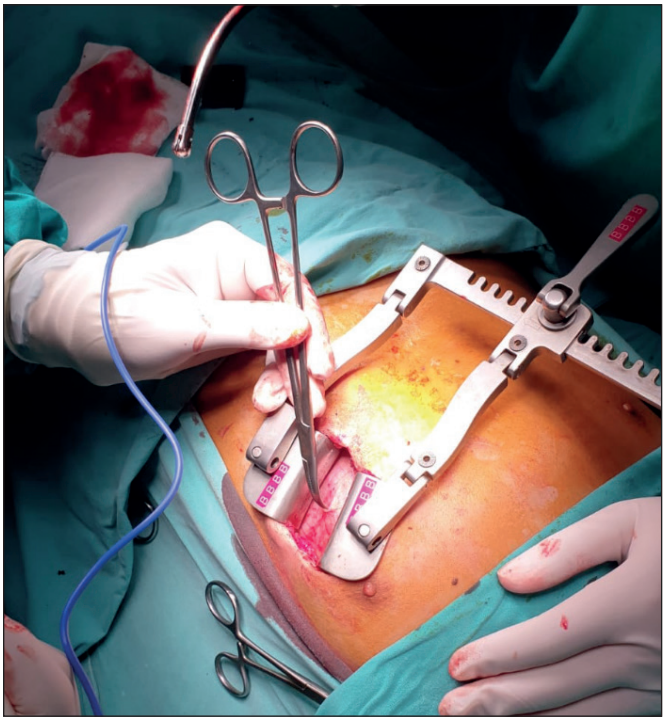

**Figure 5:** Window where the pleural effusion is drained.

At the initial phase of anesthesia, mechanical ventilator settings were set as tidal volume 6 ml/min, frequency 14/min, peak airway pressure 25 cmH2O, etCO2 30-35 mmHg, and 50% O2/air mixture. At the beginning and during the operation, mechanical ventilator tidal volume was set at 4 ml/min and frequency at 18/min, and etCO2 was regulated according to blood gas values.

A window of 1-2 cm was opened pericardially by means of a left anterior mini-thoracotomy and 1500 cc hemorrhagic fluid was drained. A pericardial biopsy specimen was taken (Figure 5). A 28 F chest tube was placed, and the surgery was successfully completed in 65 minutes. The RLDT was replaced with a single lumen intubation tube. The patient was transferred to the intensive care unit (ICU) where cardiovascular surgery patients were followed postoperatively. The patient was not sedated in intensive care. The patient reached extubation criteria and was extubated after 3 hours in the ICU. The patient was transferred to the hospital ward from the ICU on the following day.

## DISCUSSION

Proteus syndrome is a rare congenital disease characterized by excessive growth of various tissues of the body, and its diagnostic criteria have been defined by Biesecker (2). Its characteristic findings are excessive growth in all or part of the body, macrocephaly, pigmented areas on the skin, hemangiomas, lipomatous lesions and series of symptoms related to the skeletal system. Rare findings are hydronephrosis, ophthalmologic abnormalities (42%), cystic lung diseases (12-13%), convulsion, external auditory canal stenosis, mental retardation (40%), urological abnormalities (9%), and macroorchidism. PS becomes prominent in early infancy (6 to 18 months), progresses rapidly in childhood, and progression slows after puberty. Approximately 250 PS cases have been reported in the literature. The prevalence of PS is between 1:1,000,000 and 1:10,000,000, with a male/female ratio of 1.9/1.5 indicating male predominance (3).

PS patients should be preoperatively examined in detail, and precautions should be taken during and after anesthesia and the patient carefully managed. Difficult intubation and ventilation due to deformities are the leading problems that can be encountered during anesthesia. These include impairment of upper and lower incisors, restriction of neck movements, antevert nostrils, vertebral abnormalities and kyphoscoliosis (4,5).

Regional anesthesia, endotracheal intubation with fiberoptic bronchoscope under sedation and Mc Coy laryngoscope use have been reported in PS patients. Pennant et al. reported a case of facial dysmorphism that required fiberoptic intubation under inhalation anesthesia (4,5). Although neck extension was slightly limited in our case, a 35 F RLDT was successfully placed in the right lung at once and without any difficulty in intubation and ventilation. DLT placement was confirmed by flexible bronchoscopy.

In most patients with PS, an increase in adipose tissue is seen in the abdominal, pelvis, subcutaneous tissues. Lipomatous infiltration in the heart or cardiac involvement was reported in two cases in the literature. Shaw et al. presented a 20-year-old PS case with thickened echogenic interventricular septum, right ventricular hypertrophy findings in the ECG, and a mass in the apex of the right ventricle (5,6). Even though no increase in adipose tissue in the myocardium was found in PS, it could not be identified whether this finding was associated with arrhythmias, conduction defects or haemodynamic abnormalities. Further studies and research are required for this purpose (7).

In conclusion, PS is a rare congenital disease characterized with excessive tissue growth and marginal cardiac involvement and patients should be well examined and closely monitored at all stages due to difficulties in anesthesia management. The necessary measures should be taken considering the clinical findings of the patients.

**Informed Consent:** All the participants' rights were protected and written informed consents were obtained before the procedures according to the Helsinki Declaration.

**Conflict of Interest:** The authors have no conflict of interest to declare.

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