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Konjenital kalp cerrahisi sonrası dornaz alfa (pulmozyme®) ile postoperatif atelektazinin yenidoğanda başarılı tedavisi**Successful treatment of postoperative atelectasis with dornase alpha (pulmozyme®) after congenital heart surgery in a newborn**Ahmet ÖZYAZICI¹Nurdan Dinlen Fettah¹Rumeysa ÇİTLİ¹Ahmet Öktem¹Ayşegül Zenciroğlu¹ Orcid ID:0000-0002-1389-7799 Orcid ID:0000-0001-7530-1172 Orcid ID:0000-0002-0793-6608 Orcid ID:0000-0001-7209-6732 Orcid ID:0000-0002-3488-4962¹ SBÜ Dr. Sami Ulus Kadın Doğum Çocuk Sağlığı ve Hastalıkları Eğitim ve Araştırma Hastanesi, Yenidoğan Kliniği, Ankara, Türkiye**ÖZ**

On iki günlük erkek hasta suprakardiyak total anomalous pulmonovenöz bağlantı (TAPVC) ile takip edildi ve postoperatif birinci gününde servise kabul edildi. Ameliyat sonrası akciğerlerde atelektazi gelişmesi nedeniyle hastanın ekstübasyon girişimi üç kez başarısız oldu. Atelektazidornase alfa ile tedavi edildi. Hastanın takibinde yineleme atelektazisi olmadı ve oksijen gereksinimi olmadan hasta taburcu edildi.

Anahtar kelimeler: Atelektazi, dornaz alfa (pulmozim), konjenital kalp hastalığı**ABSTRACT**

12 day male patient followed with supracardiac total anomalous pulmonary venous connection (TAPVC) was admitted to our service after one day of surgical repair. The patient extubation attempts failed three times because of the atelectasis occurred in the lungs after the surgery. Atelectasis was treated with installed dornase alpha. It avoided the recurrence of atelectasis and the patient was discharged with no oxygen.

Keywords: Atelectasis, dornasealpha (pulmozyme), congenital heart surgery**INTRODUCTION**

Congenital heart disease (CHD) is associated with substantial morbidity and accounts for 4% of all neonatal deaths (1). About 25% of babies with a CHD have a critical CHD. Infants with critical CHDs generally need surgery or other procedures in their first year of life (2). Cardiac and pulmonary pathophysiologies are closely interdependent, which makes the management of patients with CHD all the more complex. Pulmonary complications of CHD can be structural due to compression causing airway malacia or atelectasis of the lung (3). Atelectasis, defined as collapse of a certain region of the lung parenchyma is the most common complication in the postoperative period of cardiac surgery by worsening oxygenation, decreasing pul-

monary compliance, leading to inhibition of cough and pulmonary clearance and may lead to respiratory failure and increase pulmonary vascular resistance (4,5). Pulmonary complications are the most common causes of morbidity and mortality in the postoperative period after congenital heart surgery (3). Recombinant human DNase (rhDNase) has proven to be an effective treatment in opening airways in cystic fibrosis (6). In infections complicated by atelectasis, bronchial secretions and mucus plugs also have a high concentration of DNA such that DNase could also be an effective treatment in this situation.

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Başvuru tarihi : 02.01.2021

Kabul tarihi :01.12.2021

CASE

12 day male patient followed with supracardiac total anomalous pulmonary venous connection was admitted to our service after one day of surgical repair. He was diagnosed with TAPVC after five days of birth with echocardiography and cardiac catheterization. On the fourth post operative day when he met extubation criteria he was extubated. After 2 hours of extubation he had difficult breathing, his arterial blood gas was analysed he had respiratory acidosis and he was re-entubated. The chest x ray showed parsiyel atelectasis of the right lung (Fig 1).

Fig .1 Parsiyel atelectasis of the right lung



Pulmonary rehabilitation techniques and bronchodilators were used for the treatment of atelectasis. rhDNase (Pulmozyme®; Roche, Basel, Switzerland) was administered either as a 2.5 mg dose nebulised twice daily. After 3 days of treatment his chest x ray remains stil the same and extubation attempts failed. We started 10% of this dose was diluted to 5 ml with NaCl 0.9% and given slowly as droplets into the endotracheal tube. This treatment was continued until the atelectasis had improved sufficiently, preferably based on the chest X-ray of the next day. This dose was chosen as it was estimated that pulmonary deposition of a regular 2.5 mg dosage would be a maximal 10%. When rhDNase was instilled endotracheally, it was attempted to position the head as favorably as possible for the DNase to reach the affected lobe(s). Pulmozyme is a recombinant human deoxyribonuclease I (rhDNase) an enzyme which selectively cleaves DNA. Each ampule has 2.5 mL of the solution. Each mL of aqueous solution contains 1 mg dornase alfa. After administration of 2.5 mg of Dornase alpha twice a day for three days the atelectasis of the right lung started to recover (Fig 2).

Fig. 2 rhDNase treatment resolved almost complete atelectasis



No adverse event was observed for both administration options of rhDNase. The patient was discharged from the Neonatal Intensive Care Unit on the 25 day of his life with no oxygen .

DISCUSSION

Atelectasis in patients with CHD can be attributed to extrinsic compression from vascular malformation, restrictive defects from pulmonary oedema or from underlying respiratory tract infection. There are many postoperative factors that place a patient at risk for the development of atelectasis. These include immobilization, splinting, cough suppression, mucus plugging and hypoventilation from pain and sedation. Atelectasis reduces lung compliance, increases work of breathing, and causes ventilation/perfusion mismatching that result in hypoxemia (7). There is no 'golden standard' for treatment of atelectasis in children. Efficacy of treatment modalities such as inhaled bronchodilators, steroids, physiotherapy and nebulised sodium chloride (NaCl 0.9%) has not been demonstrated (8).

Dornase alfa is a recombinant enzyme well known for the treatment of cystic fibrosis: it reduces viscosity of airway secretions, breaking bonds between extracellular DNA molecules derived from leukocytes and infectious agents (9). Its efficacy has been well documented in cystic fibrosis, whereas case reports have described a beneficial effect in congenital hearth disease (10-11). Dornase alfa also reduced ventilation days, while there was a trend towards shorter hospital stay and less atelectasis in congenital heart disease children (12)

In conclusion, instilled Dornase alfa has a dramatic effect in resolving atelectasis and sholud be considered in cases of persistent atelectasis due to pulmonary medication and rehabilitation program after congenital heart surgery (13, 14).

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