Acute pulmonary edema in a newborn with infracardiac type total anomalous pulmonary venous return and surgical repair

Yusuf Kenan Yalçınbaş, Ersin Erek, Ece Salihoğlu, Nilüfer Öztürk, Gökhan Mamur
Nerime Soybir, Ayşe Sarıoğlu, Tayyar Sarıoğlu
İstanbul Memorial Hospital, İstanbul, Turkey

SUMMARY: Yalçınbaş YK, Erek E, Salihoğlu E, Öztürk N, Mamur G, Soybir N, Sarıoğlu A, Sarıoğlu T. Acute pulmonary edema in a newborn with infracardiac type total anomalous pulmonary venous return and surgical repair. Turk J Pediatr 2004; 46: 179-181.

Total anomalous pulmonary venous return (TAPVR) is a rare congenital pathology. Early diagnosis and urgent surgery are life-saving, especially in newborns with pulmonary venous obstruction, which is most commonly seen with infracardiac type. A three-day-old baby boy presented to another clinic with tachypnea and cyanosis. Initial work-up aimed at ruling out persistant pulmonary hypertension, respiratory distress syndrome and pneumonia. Acute pulmonary edema then developed, and on echocardiography obstructive type infracardiac TAPVR was suspected. Cardiac catheterization was done for definitive diagnosis. Urgent surgery was undertaken and pulmonary veins were anastomozed to left atrium with posterior approach. Patient was extubated at 10th day and discharged after three weeks. During one-year follow-up the patient was free of symptoms. Infracardiac type TAPVR is a rare pathology in which early diagnosis and urgent surgery with special postoperative case are mandatory for survival.

Key words: newborn, acute pulmonary edema, infracardiac type total anomalous pulmonary venous return.

Total anomalous pulmonary venous return (TAPVR) is a rare congenital cardiac anomaly (1.5-3%) and presents with a wide spectrum of anatomical and clinical variations[1]. Prognosis depends on the degree of pulmonary venous obstruction. Twenty-five percent of cases are infracardiac type with pulmonary venous obstruction and pulmonary hypertension. Mixed types are also occasionally seen after the newborn period[2]. Urgent diagnosis and surgical intervention are life-saving in cases of acute pulmonary edema soon after birth[3,4]. In this report, medical and surgical approaches to a newborn with infracardiac type total anomalous pulmonary venous return and acute pulmonary edema are presented.

Case Report

A six-day-old baby boy was brought to our pediatric outpatient clinic due to increasing jaundice and mild difficulty in breathing. The baby was born to a 28-year-old gravida 5, para 3 mother via elective cesarean section at term. Mild jaundice and heart murmur were noted in newborn nursery and he was discharged on the 3rd day of life with instructions regarding jaundice and murmur.

On physical examination the baby was awake, mildly cyanotic and alert, but irritable in mild-to-moderate respiratory distress with tachypnea (80/min) and intercostals retractions. Bilateral diffuse fine crackles and a grade 3/6 systolic murmur at left lower sternal border were noted. Chest X-ray showed bilateral diffuse pulmonary interstitial densities. Cardiac borders were obscured. His echocardiographic examination revealed pulmonary hypertension, secundum type atrial septal defect (ASD) and mild-to-moderate degree of tricuspid insuficiency[1].

He was admitted to neonatal intensive care unit and therapy was directed towards the preliminary diagnosis of persistant pulmonary hypertension. Despite initiation of parenteral antibiotics and supplemental oxygen, his condition rapidly

deteriorated and mechanical ventilation with inotropic support was started two days after his hospitalization. His repeat chest X-ray revealed diffuse pulmonary edema, and cardiac shadow was lost despite controlled ventilation, use of inotropics and diuretics (Fig. 1). Rapid development of acidosis, hepatomegaly and generalized edema raised the question of a cardiac pathology and a repeat ECHO examination was performed. The findings were suggestive of infracardiac type TAPVR with obstruction which was then confirmed with a catheter study. All four pulmonary veins were draining into the subdiaphragmatic hepatic venous system via a vertical vein which was obstructed at the distal end (Fig. 2).

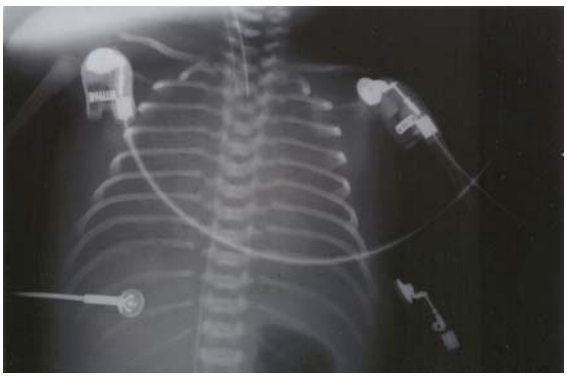

Fig. 1. Preoperative X-ray depicting diffuse pyulmonary edema to such extent that cardiac shadow is lost.

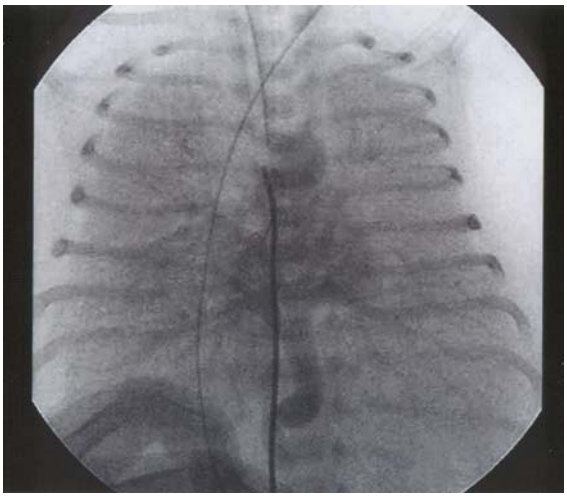

Fig. 2. Preoperative angiography. All four pulmonary veins are draining into subdiaphragmatic obstructive venous system. White arrow points out vertical vein.

The patient was transferred to the operating room and cardiopulmonary support was initiated. Pulmonary venous chamber, which was located behind the heart, was connected to the left atrium. A longitudinal incison was made on the posterior wall of the left atrium starting from the atrial appendix down to mitral annulus. Another incision was made on the corresponding site of the pulmonary venous chamber. Nonobstructive anastomosis was created between these two chambers. Atrial septal defect was closed with autologous pericardial patch and the vertical vein was ligated.

Deep sedation, paralysis, mechanical ventilation and prostaglandin were used for the management of pulmonary hypertension in the first 72 hours. Postoperative chest X-ray showed the immediate resolution of pulmonary edema (Fig. 3). Adenosine was used for the early intermittent supraventricular tachycardia. The patient was extubated on the 10th postoperative day and discharged three weeks later in good condition.

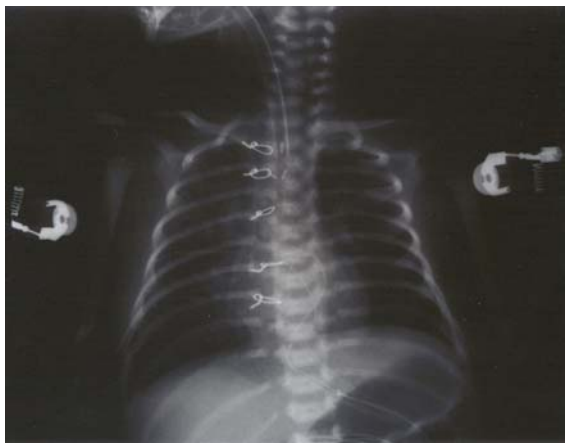

Fig. 3. Postoperative X-ray shows complete resolution of diffuse pulmonary edema immediately after repair.

Regular postoperative follow-up, including echocardiograpic examination, was done at one month and every three months thereafter. As of the last follow-up at one year of age, the baby is growing well, reaching milestones appropriately and is free of symptoms. No sign of pulmonary venous obstruction has been detected in his serial echocardiographic examinations.

Discussion

Current mortality is less than 10% for most types of TAPVR at special centers with advanced surgical and medical facilities.

However, for obstructive infracardiac type, TAPVR mortality is still high, approaching 30%[3-5]. The main reason for the poor outcomes is the nonpalliative nature of the pathology due to mechanical obstruction to pulmonary venous blood at the inferior end of the vertical vein. In fact, this pathology is accepted as the only true emergency situation in desperate need for surgery among the wide spectrum of congenital cardiac abnormalities[6,7]. Prostaglandins, balloon septostomy and mechanical ventilation are useful for most of the babies with neonatal congenital cardiac abnormalities awaiting early surgical intervention (transposition of the great arteries, tricuspid atresia, hypoplastic left heart variants, etc). In our case, urgent anastomosis of the venous chamber with the left atrium relieved diffuse pulmonary edema immediately after surgery and could not have been achieved by other available treatment options.

Total anomalous pulmonary venous return with pulmonary venous obstruction almost always presents with severe symptoms in the early hours or days of life[6]. Pulmonary edema and difficulty in breathing should raise the question of this pathology along with other differential diagnoses such as respiratory distress syndrome and bronchopneumonia[3]. Echocardiography is very helpful in establishing diagnosis but occasionally a catheter study is needed for confirmation. It is probably best to avoid osmotic load due to contrast agents in critical newborns if echocardiographic views are conclusive.

Surgical technique is aimed to create a posterior non-obstructive pulmonary venous chamber and left atrium connection with a wide circumferential anastomosis[11,12]. ASD is closed with a patch and vertical vein is either ligated or left alone[3,4]. Some authors prefer not to ligate or gradually close the vertical vein with adjustable sutures with the idea that it would decompress the heart in the early postoperative period and reduce mortality and morbidity[9,10].

Since the early postoperative mortality is mostly related to pulmonary hypertensive crisis, the management must be geared towards avoidance of such events. Deep sedation, paralysis, and hyperventilation to achieve 100% O_2 saturation and low normal CO_2 have to be maintained[7]. We routinely manage infants with pulmonary hypertension with the above measures and start weaning once pulmonary pressures stabilize under mild sedation.

On long-term follow-up pulmonary venous obstruction might develop. Usually obstruction is at the level of anastomosis but occasionally diffuse stenosis at the level of intrapulmonary veins might occur[5,8]. In our case, over the one-year follow-up, there was no detectable pulmonary venous obstruction on echocardiography.

In conclusion, infracardiac type TAPVR is a rare and high-risk congenital cardiac abnormality that needs early diagnosis and surgical treatment with special care in the intensive care unit. Successful results are obtained with comprehensive multidisciplinary team effort in specialized centers.

REFERENCES

1. Sarıoğlu T, Özme Ş, Oto Ö, et al. Total anomalous pulmonary venous drainage-surgical correction. Turk J Pediatr 1985; 27: 69-77.
2. Sarıoğlu T, Kınaoğlu B, Paker T, Sarıoğlu A, Saltık L, Aytaç A. A rare case of mixed type total anomalous pulmonary venous connection and its surgical treatment. Thorac Cardiovasc Surg 1997; 45: 152-154.
3. Barrat-Boyes BG, Kirklin JW. Total anomalous pulmonary venous connection. In. Cardiac Surgery (2nd ed.) Vol.1. New York: Churchill Livingstone Inc; 1993: 645-673.
4. Castenada AR, Jonas RA, Mayer JE, Hanley FL. Anomalies of the pulmonary veins. In: Cardiac Surgery of the Neonate and Infant. Philadelphia, Pennsylvania: W.B. Saunders Co; 1994: 157-166.
5. Hyde JA, Stümper O, Barth MJ, et al. Total anomalous pulmonary venous connection: outcome and management of recurrent venous obstruction. Eur J Cardiothorac Surg 1999; 15: 735-740.
6. von Son JA, Hambsch J, Kinzel P, et al. Urgency of operation in infracardiac total anomalous pulmonary venous connection. Ann Thorac Surg 2000; 70: 128-130.
7. Sinzobahamvya N, Arenz C, Brecher AM, et al. Early and long-term results for correction of total anomalous pulmonary venous drainage (TAPVD) in neonates and infants. Eur J Cardiothorac Surg 1996; 10: 433-438.
8. Caldarone CA, Najm HK, Kadletz M, et al. Relentless pulmonary vein stenosis after repair of total anomalous pulmonary venous drainage. Ann Thorac Surg 1998; 66: 1514-1520.
9. Caspi J, Pettitt TW, Fontenot EE, et al. The beneficial hemodynamic effect of selective patent vertical vein following repair of total anomalous pulmonary venous drainage in infants. Eur J Cardiothorac Surg 2001; 20: 830-834.
10. Caspi J, Zalstein E, Zuker N, Appelbaum A. Adjustable suture of the vertical vein for repair of infracardiac total anomalous pulmonary venous connection. J Thorac Cardiovasc Surg 1995; 109: 389-390.