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*Brief Report*

# Changes in 99mTechnegas ventilation lung scan in a newborn with absent pulmonary valve syndrome

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**Abstract** A newborn infant with tetralogy of Fallot and absent pulmonary valve was successfully corrected in two stages. Absent pulmonary valve syndrome presenting in early infancy manifests severe respiratory symptoms that still make challenging both management and surgical treatment. This is ascribed to tracheobronchial compression by the extremely dilated pulmonary arteries, and to the resultant pulmonary obstructive lesions. We report herein the first findings of 99mTechnegas ventilation lung scanning in an infant with the syndrome to assess the pulmonary obstructive lesions.

Keywords: Congenital heart disease; pulmonary obstructive lesion; scintigraphy

Severely symptomatic neonates with the absent pulmonary valve syndrome usually suffer high mortality and morbidity. The surgical options that have proved successful in older infants still present many challenges in these cases with early onset.[1] The respiratory symptoms are ascribed to tracheobronchial compression by the extremely dilated pulmonary arteries, and to the resultant pulmonary obstructive lesions.[2] To the best of our knowledge, we have employed, for the first time, 99mTechnegas ventilation and perfusion lung scanning to assess the pulmonary obstructive lesions in an infant with the syndrome. We now describe the clinical link between the pulmonary obstructive lesions and the postoperative respiratory failure.

## Case report

The baby boy was born at 41 weeks gestation, weighing 2,726 g, with profound cyanosis and severe respiratory distress that prompted endotracheal intubation soon after the birth. On referral to our institute, he had developed bilateral pneumothorax that required drainage by chest tubes as an emergency. Successful intensive care consisted of mechanical ventilatory support, administration of inotropes, inhalation of nitric oxide, and so on. Despite the relief of hypoxemia, hypercarbia with carbon dioxide tension in excess of 90 mmHg was persistent. Acceptable ventilation could be achieved only with the infant in the lateral decubitus position. We also noted that hypercarbia was exacerbated with the use of muscle relaxants.

The chest X-ray showed cardiomegaly with dilated pulmonary arteries and hyperinflated right lower lung. The diagnosis of tetralogy of Fallot with absent pulmonary valve was made on ultrasonic examination. Preoperative cardiac catheterization was performed. Angiography confirmed the diagnosis, demonstrating aneurysmal dilation of the pulmonary arteries and massive pulmonary regurgitation. The ratio of pulmonary-to-systemic flow was 1.8. The pulmonary arterial index described by Nakata et al.[3] was tremendously high at 1,502 $mm^2$/body surface area. Initial palliation, at the age of 12 days, consisted of tight banding of the pulmonary trunk, and a 4 mm modified left-sided Blalock-Taussig shunt was constructed. He could not bear pulmonary ligation due to severe intraoperative hypoxia and hypercarbia. The postoperative respiratory management required mechanical ventilatory support for one month in the lateral decubitus position. Postoperative cardiac catheterization revealed the pressure in the pulmonary arteries to be 18/12(17), with the pressure in the right ventricle measured at

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95/end10, the ratio of pulmonary-to-systemic flow calculated at 0.9, and the pulmonary arterial index at 1747. Concomitant bronchography disclosed diffuse areas of narrowing in the secondary branches of the tracheobronchial tree, especially in the right lung (Fig. 1).

He was eventually discharged with an acceptable arterial oxygen saturation of 84% at the age of two months. He had developed no significant airway infection. His psychomotor development was intact. Cardiac catheterization done six months after the palliation revealed the pressure in the pulmonary arteries to be 16/14(15), that in the right ventricle of 92/end10, a ratio of pulmonary-to-systemic flow of 0.8, and the pulmonary arterial index at 1738.

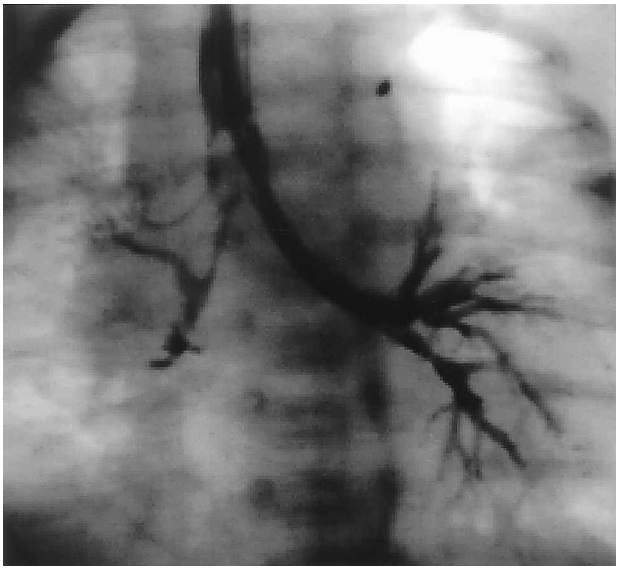

**Figure 1.**
*Bronchography in antero-posterior position performed after banding the pulmonary trunk. Diffuse areas of narrowing can been seen in the secondary branches of the tracheobronchial tree: complete occlusion of the right middle bronchiole and partial occlusion of the right lower and left upper bronchioles.*

Perfusion lung scanning with 99mTc macroaggregated albumin showed the presence of a perfusion defect in the right lung. 99mTechnegas ventilation lung scanning disclosed the presence of a hypoventilation area in the same segment.

At the age of one year, he underwent radical repair, which consisted of reconstruction of the right ventricular outflow tract by a transannular patch with a monocusp, closure of the ventricular septal defect, plication of the right pulmonary artery, and creation of a posterior leaflet in the pulmonary trunk using the resected right pulmonary arterial wall. The postoperative course was complicated by difficulties with mechanical ventilation as well as with hemodynamic compromise for as long as two weeks. Even after attainment of hemodynamic stability, bronchospasms prolonged the need for mechanical ventilation with the use of inhalational bronchodilators. The ventilatory setting aimed for minimum pulmonary barotrauma. Five months after the surgical correction, he was extubated and clinically stable on the regimen of oral bronchodilators and diuretics. His arterial oxygen saturation was 90% in room air. Cardiac catheterization done nine months after the repair revealed pressures in the pulmonary arteries of 35/3(18), pressure in the right ventricle at 40/end10, the ratio of pulmonary-to-systemic flows of 1.0, and the pulmonary arterial index at 2045. Under 100% oxygen inhalation, the pulmonary arterial pressure decreased to 30/4(15) and his arterial oxygen saturation increased to 100%. The pulmonary obstructive lesions were persistent judging from the ventilation and perfusion lung scans (Fig. 2).

## Discussion

A newborn infant with tetralogy of Fallot and absent pulmonary valve was successfully corrected with two-stage repair. The postoperative respiratory failure subsided with time after ventilation in the lateral decubitus position, and ended in successful

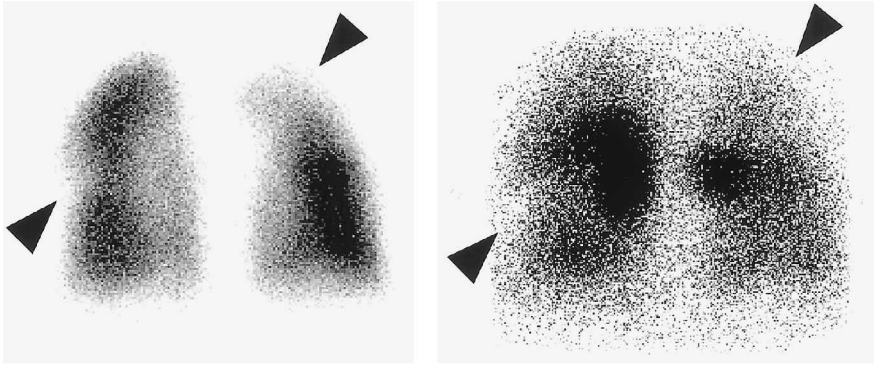

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**Figure 2.**
*(a) Perfusion lung scan with 99mTc macroaggregated albumin after radical repair. Arrowheads mark hypoperfusion areas. (b) 99mTechnegas ventilation lung scan after radical repair. Arrowheads mark hypoventilation areas.*

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extubation one month after the palliation. Cardiac catheterization revealed no reduction of the pulmonary arterial index, but decreased pulmonary regurgitation. On the other hand, he developed nearly intractable ventilatory failure and hemodynamic compromise after the radical repair. Even after hemodynamic stability was attained, quite unstable bronchospasms prolonged the need for mechanical ventilation for 5 months.

The respiratory failure associated with absent pulmonary valve syndrome results from bronchomalacia due to severe tracheobronchial compression by the dilated pulmonary arteries, and from pulmonary obstructive lesions.[2] To assess the severity of the pulmonary obstructive lesions, we performed perfusion lung scanning with 99mTc macroaggregated albumin, and 99mTechnegas ventilation lung scanning, both of which have proved to be diagnostic for pulmonary obstructive lesions such as chronic obstructive lung disease.[4] 99mTechnegas ventilation lung scanning definitely demonstrated hypoventilated area in his lungs corresponding to the hypoperfusion areas seen on the perfusion lung scan. This pulmonary obstructive lesion, verified by bronchography, may have resulted in severe respiratory failure and hemodynamic compromise. When ventilation and perfusion lung scans disclose prominent obstructive lesions, reconstruction of the right ventricular outflow tract using a homograft[5] might have been an option for reducing pulmonary regurgitation.

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