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Journal homepage: www.ijca.in **Brief Communication****A rare case of congenital bilateral bullous emphysema with vacterl anomaly with congenital heart disease****Pritam Jadhav^{1,*}, Maheshrao Thorat¹, Nikhil Shetty¹**¹Dept. of Pediatric Cardiac Anaesthesiology and Intensive Care, Jupiter Hospital, Thane, Maharashtra, India **ARTICLE INFO***Article history:*

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Dear editor,

Congenital bullous emphysema is defined as large bullae involving one third of hemithorax.¹ A 2yr, 9 months old female weighing 8.2kg presented with dyspnea on playing was posted for surgical correction of congenital heart disease. She is a case of VACTERL anomaly having polydactyly, vertebral anomalies and single right kidney, her developmental milestones was delayed and immunisation was complete. On 2D echocardiography she was diagnosed with ventricular septal defect with multiple atrial septal defect with severe pulmonary arterial hypertension. Routine investigation were within normal limits. Chest radiography showed right sided pneumothorax so we decided to insert intercostal drain and proceed with diagnostic catheter study for evaluation for severe pulmonary arterial hypertension. Chest radiography revealed no reduction in pneumothorax so we did high resolution computed tomography which showed bilateral emphysema involving middle and lower lobe of both lung (Figure 1). The radiographic criteria for congenital bullous emphysema as defined by Roberts and colleagues, include the presence of giant bullae in one or both upper lobe, middle lobe, lower lobe, occupying atleast one third of hemithorax and compressing surrounding normal lung parenchyma.² Differential diagnosis include bronchial atresia, bronchogenic cyst, Swyer-james syndrome and congenital cystic adenomatoid

malformation. Surgical removal of affected lobe is commonly done. We do conservatively manage some patients who are not clinically in respiratory distress and able to feed and grow. Maintaining ventilator pressures and volume as low as possible avoids producing ventilator related hyper-expansion of the affected lobe. Management by more conservative, gentle ventilation techniques if successful will result in fewer emergency surgeries with congenital lobar emphysema.³

Fig. 1: High resolution computed tomography showing bullous changes and progressive emphysema and intercostal drain in position

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For performing emergency lobectomy the anaesthetic management would be a challenge. After induction, PEEP

can expand the affected lobe and this can compress the normal lung resulting in cardiovascular compromise. Once resection of the affected lobe is completed controlled lung ventilation with muscle relaxation should be done. Blood gas monitoring intraoperatively and postoperatively should be done. Intercostal block is a good for pain control. Lung biopsy of the resected part should be done.

Conflicts of Interest

There are no conflicts of interest.

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