Congenital bronchial atresia with regional emphysema associated with pectus excavatum

R J van Klaveren, W J Morshuis, L K Lacquet, A L Cox, J Festen, F M J Heystraen

Abstract
Two cases of congenital bronchial atresia with pectus excavatum are reported. Costosternal retraction during the efforts to overcome the airway obstruction due to encroachment on normal lung tissue by the hyperinflated segments may play a part in causing pectus excavatum.

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Congenital bronchial atresia, representing one cause of congenital regional emphysema, is a rare anomaly. Since the first description by Ramsay and Byron in 1953, 87 cases have been reported. 2 To our knowledge this is the first report of congenital bronchial atresia associated with pectus excavatum. We discuss the possible cause and effect relationship and the indications for surgical treatment.

Case reports
Patient 1
A 23 year old woman was admitted for correction of pectus excavatum in 1980. She had had recurrent bronchopulmonary infections, dyspnoea, chest pain, and psychological problems associated with her pectus excavatum. Physical examination showed an asymmetric pectus excavatum with the deepest impression on the right side. On the chest radiograph there was loss of normal lung markings in the right upper and mid zones and a low grade opacity mass lesion in the area of the right hilum (figure). Bronchography and bronchoscopy showed atresia of the anterior segmental bronchus of the right upper lobe. Pulmonary angiography showed downward displacement of the middle and lower lobe arteries due to emphysema in the upper lobe. For psychological reasons correction of the pectus deformity was performed. In the same session the hyperinflated right anterior segment and posterior subsegment were resected to prevent further bronchopulmonary infections. Examination of the specimen showed a proximal blind ending bronchus with a mucocoele. Distally very small bronchi up to 3 mm in diameter were seen to be connected with emphysematous, deformed alveoli. Some of the distal bronchi did not contain cartilage.
been reported before. An increased negative intrathoracic pressure during inspiration to overcome airway obstruction has been suggested as resulting in costal sternal retraction. It was found that relief of airway obstruction by tonsillectomy or surgical treatment of laryngomalacia resulted in resolution of pectus excavatum. In asthmatic children the prevalence of pectus excavatum was found to be two to three times greater than even the highest prevalence in the normal population. Godfrey reported four children with pectus excavatum and a collapsible segment in the main bronchus due to lack of cartilage support. A relationship between pectus excavatum and congenital regional emphysema was suggested by Shaw. Whether pectus excavatum can be solely ascribed to an underlying lung abnormality is, however, questionable. Overgrowth of the costal cartilage has been generally accepted as a causative mechanism of pectus excavatum. Decreased mechanical stability as a result of age dependent morphological and biochemical changes found in costal cartilage does account for the progression of asymmetry with increasing age. In patients with congenital bronchial atresia costosternal retraction during the efforts to overcome airway obstruction due to encroachment of the hyperinflated segments on normal lung tissue may aggravate the natural course of pectus excavatum. This theory might be supported by the fact that the side of the deeper impression corresponds with the side of the congenital bronchial atresia.

When congenital bronchial atresia gives rise to symptoms (recurrent bronchopulmonary infections, dyspnoea, chest pain) surgical treatment should be undertaken. The treatment of symptomless patients, however, is controversial. Because 85% of them have been operated on in the past, the natural history is unknown. Recurrent bronchopulmonary infections, pneumothorax, or encroachment on normal lung tissue by the hyperinflated lung might occur and justify surgical excision to avoid such complications in future. When pectus excavatum is present we advise operating on congenital bronchial atresia at the same session. The optimal age for operating on pectus excavatum is 5–10 years. Careful examination of the routine chest radiograph should be performed in patients with pectus excavatum to exclude the possible presence of congenital bronchial atresia.

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