

Pneumothorax And Bullae In Marfan Syndrome

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Pneumothorax And Bullae In Marfan

Background: Increased risk of spontaneous pneumothorax has been described in patients with Marfan syndrome and has been attributed, in part, to the presence of apical blebs and bullae. Objectives: We assess the risk of pneumothorax and its relationship to the presence of apical blebs and bullae in patients with Marfan syndrome in the era of CT imaging.

Pneumothorax and bullae in Marfan syndrome

pneumothorax compared to 4 of 150 patients (2.7%) without blebs or bullae (p = 0.003). CONCLUSIONS: The frequency of blebs is relatively low in patients with Marfan syndrome but the risk of pneumothorax is significantly higher in those with radiologically detectable blebs or bullae. Chest CT scanning to identify blebs

Pneumothorax and bullae in Marfan syndrome.

Conclusions: The frequency of blebs is relatively low in patients with Marfan syndrome but the risk of pneumothorax is significantly higher in those with radiologically detectable blebs or bullae. Chest CT scanning to identify blebs and bullae may allow risk stratification for pneumothorax in patients with Marfan syndrome. KW - Bullous diseases

Pneumothorax and bullae in Marfan syndrome — Mayo Clinic

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Pneumothorax and Bullae in Marfan Syndrome

Although pulmonary symptoms are not generally considered a main feature of Marfan syndrome, many patients have a degree of underlying pulmonary pathology, such as cystic changes, emphysema, spontaneous pneumothorax (SP), focal pneumonia, bronchiectasis, bullae, congenital pulmonary malformations, and apical fibrosis [2, 3].

Thorascopic Treatment of Pneumothorax in Marfan Syndrome ...

Six patients had new mutations of the Marfan syndrome. Spontaneous pneumothorax was bilateral or recurrent in 7 patients. Nine patients had underlying lung disease, in the form of diffuse or apical bullae, visible on chest radiographs. Three patients had severe pectus excavatum and 2, severe pedus carinatum.

Pneumothorax in the Marfan Syndrome: Prevalence and ...

The increased risk of pneumothorax has been attributed to the presence of apical blebs, bullae, abnormal connective tissue constituents in the lung parenchyma or increased mechanical stresses in the lung apices due to the tall body habitus. 2 The causal gene for Marfan's syndrome, FBN1, encodes the extracellular matrix glycoprotein fibrillin-1, which can be found in the lung as a component of elastic fibres, it has been proposed that abnormalities of fibrillin result in connective tissue ...

Spontaneous pneumothorax as manifestation of Marfan syndrome

In the Marfan lung, pneumothorax can be recurrent, present in both lungs, and associated with emphysema. A person with pneumothorax may experience shortness of breath, dry cough, an acute onset of pleuritic chest pain (chest pain that gets worse when you take a deep breath), or chest pain that gets worse when coughing.

Lungs | The Marfan Foundation

Distinction of pneumothorax from emphysematous bullae The bullae of emphysema can be very large and, when situated in the periphery of the lung, can mimic a loculated pneumothorax. A chest drain inserted into a bulla in the mistaken belief that it is a pneumothorax is not uncommon.

Air leaks, pneumothorax, and chest drains | BJA Education ...

patients, apical bullae were present and detectable on chest radiograph. Pneumothorax frequently recurred if not treated by resection of the offending bullae. We recom- mend that all adolescent and adult patients with the Marfan syndrome undergo chest radiography for ascertain-

Pneumothorax in the Marfan Syndrome: Prevalence and Therapy

Pneumothorax is one of the criteria of the Marfan systemic score (80). Among eight patients, the median number of pneumothoraces was one (range, one to three) (83). Blebs or bullae are a risk factor for pneumothorax; pectus excavatum and smoking are not (83).

The Genetics of Pneumothorax | American Journal of ...

Smoking, and smoking cannabis have been implicated in the development of spontaneous pneumothorax in young (otherwise healthy) patients. Bullae, or air pockets within the lung tissue are more commonly associated with chronic disease processes such as chronic obstructive pulmonary disease (emphysema).

Blebs, Bullae and Spontaneous Pneumothorax - Thoracic Surgery

6. Distinguish between a large bulla and a pneumothorax Chest radiograph shows unilateral hyperlucency affecting the entire right lung (white arrows). (b) CT demonstrates a large bulla (star) on the right side causing considerable compression of the mediastinum Investigations - CT scanning 39. CT shows large asymmetric upper lobe bullae.

Pneumothorax and pneumomediastinum

Giant bullous emphysema (GBE), otherwise known as vanishing lung syndrome, is defined as bullae occupying at least one-third of the hemithorax of one or both lungs. GBE is typically described in young male smokers. Other less common associations, including HIV infection, marijuana inhalation, and illicit injection drug use have also been reported.

Marfan Syndrome Presenting as Giant Bullous Emphysema ...

Early diagnosis based on untypical symptoms of Marfan syndrome like pneumothorax and bullae helped in the early evaluation of cardiovascular system and early managements of the disease, which proved to increase life expectancy of patients with Marfan syndrome markedly.

Marfan syndrome with pneumothorax: case report and review ...

Rigante D, Segni G, Bush A. Persistent spontaneous pneumothorax in an adolescent with Marfan's syndrome and pulmonary bullous dysplasia. Respiration 2001; 68:621. Hoag JB, Sherman M, Fasihuddin Q, Lund ME. A comprehensive review of spontaneous pneumothorax complicating sarcoma. Chest 2010; 138:510. Karpman C, Aughenbaugh GL, Ryu JH.

Pneumothorax in adults: Epidemiology and etiology - UpToDate

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