

# Pneumothorax And Bullae In Marfan Syndrome

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Pneumothorax and bullae in Marfan syndrome 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 Marfa ...

Pneumothorax and bullae in Marfan syndrome

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 — Mayo Clinic

Increased risk of spontaneous pneumothorax has been described in patients with Marfan syndrome and has been attributed, in part, to

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the presence of apical blebs and bullae. We assess the risk of...

### Pneumothorax and Bullae in Marfan Syndrome

Bullae can arise in adolescents and young adults for a variety of reasons: abnormal destruction of lung tissue, as in deficiency of atantitrypsin or in association with chronic pulmonary infection; abnormal stresses exerted on lung segments, as 503 Hall et al: Pneumothorax in the Marfan Syndrome suggested in patients with long [33] or deformed [39] chest cavities; congenital defects; and abnormal integrity of the connective tissue framework of the lung.

### Pneumothorax in the Marfan Syndrome: Prevalence and ...

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### [PDF] Pneumothorax And Bullae In Marfan Syndrome

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 ...

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

### Thoracoscopic Treatment of Pneumothorax in Marfan Syndrome ...

the Marfan syndrome on the occasion of his first pneumothorax. absence of breath sounds on the left and a marked in- crease in the intensity of the heart sounds.

### Pneumothorax in the Marfan Syndrome: Prevalence and Therapy

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### Pneumothorax And Bullae In Marfan Syndrome

In the Marfan lung, pneumothorax can be recurrent, present in both lungs, and associated with emphysema. A person with pneumothorax

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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. Restrictive Lung Disease

Lungs | The Marfan Foundation

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Pneumothorax and bullae in Marfan syndrome. - Abstract ...

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.<sup>2</sup> 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 ...

Pneumothorax and bullae in Marfan syndrome — Mayo Clinic 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

Pneumothorax And Bullae In Marfan Syndrome

reported in 27 and bullae with or without emphysema in 20. Although the total number of cases of Marfan syndrome "at risk" is not known, the age of the patients at least suggests a true association between the lung lesions and the Marfan syndrome. Two of our patients had aspergillomas and these may well have formed within pre-existing bullous

Pulmonary disease in patients with Marfan syndrome

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 ...

Recurrent pneumothorax is the most common complication after an initial episode and was documented in about 50% of children. 16 Primary spontaneous pneumothorax recurrence rates were highest (22.4% to 36.8%) in children 13 to 18 years of age. 5 In a retrospective study of conservative treatment for 114 Korean children with primary spontaneous pneumothorax, almost half developed ipsilateral ...

Spontaneous pneumothorax in children | The College of ...

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Studies have also shown that 5%–11% of patients with Marfan syndrome develop spontaneous pneumothorax caused by rib cage abnormalities or apical bullae due to fibrillin abnormalities that cause weakening of the connective tissue in the lungs.<sup>2 3</sup> In addition, Saita et al<sup>4</sup> observed that patients with PSP have asymmetric and flat thoraces—a clinical feature also present in most patients with Marfan syndrome (pectus excavatum), which predisposes them to the formation of subpleural bullae.

Primary spontaneous pneumothorax in conjunction with ...

A pneumothorax is an abnormal collection of air in the pleural space between the lung and the chest wall. Symptoms typically include sudden onset of sharp, one-sided chest pain and shortness of breath. In a minority of cases, a one-way valve is formed by an area of damaged tissue, and the amount of air in the space between chest wall and lungs increases; this is called a tension pneumothorax.

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