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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 ...

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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

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for pneumothorax in
patients with Marfan
syndrome. KW -
Bullous diseases

**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 the presence of

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

Pneumothorax and Bullae 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,

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Marfan Syndrome
visible on chest
radiographs. Three
patients had severe
pectus excavatum and
2, severe pedus
carinatum.

Pneumothorax in the Marfan Syndrome: Prevalence and ...

Although pulmonary
symptoms are not
generally considered a
main feature of Marfan
syndrome, many
patients have a degree
of underlying

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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 ...

Marfan syndrome was diagnosed and David I

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operation and bullectomy were performed. This case indicates that pulmonary symptoms like secondary spontaneous pneumothorax, bullae, emphysema can...

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

patients, apical bullae were present and detectable on chest

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radiograph.

Pneumothorax frequently recurred if not treated by resection of the offending bullae. We recommend that all adolescent and adult patients with the Marfan syndrome undergo chest radiography for ascertain-

Pneumothorax in the Marfan Syndrome: Prevalence and

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The cases reviewed together with other published evidence suggest that spontaneous pneumothorax and bullae are causally related to Marfan syndrome. The presence of idiopathic upper lobe fibrosis in four Marfan patients is interesting but provides insufficient evidence to assess possible causality.

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**Pulmonary disease
in patients with
Marfan syndrome.**

Beshay et al reported the largest case series of spontaneous pneumothorax in patients who frequently smoked marijuana and found severe bullous lung disease in the upper lobes of the lungs among marijuana smokers and hence recommended

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Marfan Syndrome
considering marijuana
abuse as a cause of
emphysema in young
patients.

Cannabis-induced bullous lung disease leading to pneumothorax

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

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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

Distinction of
pneumothorax from
emphysematous bullae

The bullae of

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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 ...

Pneumothorax is one of

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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

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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,

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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 usually occurred after the diagnosis in most of the cases of Marfan

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syndrome. Cases with pneumothorax as initial diagnosis of Marfan syndrome are uncommon, and even less common in adults.

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

Smoking, and smoking cannabis have been implicated in the development of spontaneous pneumothorax in

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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

Marfan's syndrome is a rare, but well defined,

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hereditary disorder with skeletal, cardiovascular, and ocular abnormalities. Pulmonary abnormalities occur infrequently in this syndrome, and they include congenital malformations, cystic disease, bullae, emphysema, spontaneous pneumothorax, and bronchiectasis. 1

Bronchiectasis and
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**Spontaneous
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Marfan's ...**

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bullae in Marfan
syndrome. The
frequency of blebs is
relatively low in
patients with Marfan
Pneumothorax And
Bullae In Marfan
Syndrome In the
Marfan lung,
pneumothorax can be
recurrent, present in

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both lungs, and
associated with
emphysema.

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These include
widespread or patchy
cystic changes,
emphysema, and
spontaneous
pneumothorax; focal
pneumonia or
bronchiectasis, bullae,
congenital pulmonary
malformations

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(particularly middle lobe hypoplasia), and apical fibrosis have also been described. 5, 11-25 However, no literature exists that has reported a critical microscopic examination of these tissues for histologic similarities. As medical and surgical treatments continue to improve, patients with Marfan syndrome are living ...

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