

# Clinical characteristics of 50 children with azygos lobe: a retrospective study

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## Research

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# Abstract

## Background

An azygos lobe is a rare anatomic variant of the lung which may be misdiagnosed as other pathological conditions. This study aimed to summarize the clinical characteristics and imaging manifestations of azygos lobe in children, which will be helpful for reasonable management and counseling of their families. Children with imaging findings of azygos lobe from West China Second University Hospital of Sichuan University (Chengdu, China) between November 2008 and February 2019 were enrolled in the study. Clinical and radiologic characteristics of these patients were collected and analyzed.

## Results

In total, 50 patients were included. 28% children were diagnosed as azygos lobe incidentally. The other 72% cases were diagnosed when they manifested respiratory symptoms including cough (18/50, 36%), fever (17/50, 34%), wheezing (9/50, 18%), dyspnea (7/50, 14%), and cyanosis (4/50, 8%). Congenital heart disease, Down syndrome, and other respiratory malformations were also found in a small proportion of these patients. In our study, all cases with azygos lobe had the right lung involved. Other imaging manifestations of these cases consisted of increased marking and infiltrates in the lung fields, local emphysema, consolidation, and atelectasis et al. Among the patients, only 8 patients (16%) diagnosed as azygos lobe infection. Compared with the non-infected group, there were no valuable risk factors related to the azygos lobe infection. And all the children with azygos lobe infection were cured after reasonable anti-infective treatment.

## Conclusion

Children with azygos lobe were diagnosed mostly because of respiratory symptoms and these children had a good prognosis after reasonable treatment. Only a small number of patients suffered from azygos lobe infection. Some of the patients with azygos lobe were found to have other organ malformations as well, which needed further monitoring and study.

## Introduction

An azygos lobe is a congenital variation of the lung. It is present in 0.2–1.2% of the population. It can mimic various pathological conditions, which presents challenges during thoracic surgical interventions. For instance, an azygos lobe may be confused with a pathological air space such as a bulla, abscess or localized pneumothorax. A consolidated azygos lobe may be confused with a neoplasm, and the abnormally located azygos vein may be mistaken for a pulmonary nodule. In order to prevent misdiagnosis and unnecessary interventions, an understanding of the clinical and imaging features of the azygos lobe is important. Here, we describe a group of children with azygos lobe.

# Materials And Methods

## Subjects and ethics statement

This study was conducted retrospectively at West China Second University Hospital, Sichuan University, a tertiary medical center in Sichuan province. Patients with imaging findings of azygos lobe between November 2008 and February 2019 were enrolled in the study. Azygos lobe was identified as the “inverted comma or tear drop shape” on chest X-ray, and a fine, convex line in the para mediastinal portion of the upper lobe on CT scan. The Institutional Review Board/Ethics Committee affiliated with West China Second University Hospital, Sichuan University, approved this study, which was performed in accordance with the ethical standards of the Declaration of Helsinki.

## Data collection and statistical analysis

Children's age, sex, manifestations, diagnosis, radiologic characteristics and other clinical information were collected. Data were analyzed using the SPSS 19.0 software package (IBM, Armonk, NY). Continuous variables were compared using Student t test or the nonparametric Mann-Whitney U test, categorical variables were compared using the chi-squared ( $\chi^2$ ) or Fisher's exact test. Two-sided P values of  $< 0.05$  were considered statistically significant.

## Results

In our study, there were 50 children diagnosed as azygos lobe on imaging from November 2008 to February 2019 in West China Second University Hospital of Sichuan University. The age of these 50 patients varied from 64-day-old to 15-year-old. The male-female ratio was 27:23. Among these patients, 28% (14/50) patients were diagnosed as azygos lobe incidentally, without any respiratory symptoms. The reasons for the chest radiological examination of the 14 children included assessment of nephroblastoma metastasis, routine screening for tuberculosis and health physical examination. The other 72% (36/50) patients were diagnosed by chest radiology when they manifested respiratory symptoms. The respiratory symptoms consisted mostly of cough (18/50, 36%), fever (17/50, 34%), wheezing (9/50, 18%), dyspnea (7/50, 14%), and cyanosis (4/50, 8%). What is more, 16% (8/50) of these patients had a history of recurrent respiratory infections (RRTIs). 12% (6/50) of patients were found to have other respiratory malformations, such as tracheal bronchus, tracheal stenosis, which were diagnosed by bronchoscopy. Some of these patients also suffered from other complications including congenital heart disease (3/50, 6%), Down syndrome (2/50, 4%). Interestingly, 4% (2/50) had a history of preterm birth (Table 1). Notably, 20 cases (20/50, 40%) with azygos lobes were not diagnosed by the first imaging examination, in other words, the rate of missed diagnosis was high.

Table 1  
Clinical features of the patients with azygos lobe

Clinical features	n(%)
Male: female	27:23
Age (year)	2 (0.17-15)*
Without respiratory symptoms	14 (28)
Assessment of nephroblastoma metastasis	1 (2)
Routine screening for tuberculosis before using corticosteroid	10(20)
Health physical examination	3 (6)
Respiratory symptoms	41 (82)
Cough	18 (36)
Fever	17 (34)
Wheezing	9 (18)
Dyspnea	7 (14)
Cyanosis	4 (8)
Recurrent respiratory infection	8 (16)
Premature	2 (4)
Infection of azygos lobe	8 (16)
Other respiratory malformations	6 (12)
Tracheal stenosis	4 (8)
Tracheal bronchus	1 (2)
Tracheal bronchus and stenosis	1 (2)
Congenital heart disease	3 (6)
Ventricular septal defect	1 (2)
Tetralogy of Fallot	1 (2)
Coarctation of aorta	1 (2)
Down syndrome	2 (4)
Missed diagnosis in the first radiology examination	20(40)
n = 50 *range	

Table 2  
Other imaging findings co-existing with azygos lobe

Complications on image	n(%)
Patchy opacity	29 (58)
Within azygos lobe	7 (14)
Within other lobes	22(44)
Emphysema	6 (12)
Within azygos lobe	1 (2)
Within other lobes	5 (10)
Consolidation	5 (10)
Within azygos lobe	1 (2)
Within other lobes	4 (8)
Atelectasis	2 (4)
Cystic adenomatoid malformation	1 (2)
Pleuritis	2 (4)
Pleural effusion	2 (4)
n = 50.	

Table 3  
Comparison of clinical features in children with or without infection in azygos lobe

Clinical feature	with infection (n = 7)	without infection (n = 43)	P
Male	5(71.40%)	22(51.16%)	0.43
Age	1(0.42-13)	2(0.17-15)	0.45
Other respiratory malformations	2(28.6%)	4(9.30%)	0.192
Recurrent respiratory infection	2(28.6%)	6(14.00%)	0.31
Congenital heart disease	0(0%)	3(6.7%)	0.47

In our study, all case with azygos lobe had the right lung involved. 54% (27/50) of patients had imaging manifestations compatible with pneumonia such as increased marking and infiltrates in the lung fields. 12%, 10% and 4% of the patients presented with local emphysema, lung consolidation, and atelectasis on image, respectively. Furthermore, a small portion of patients were diagnosed as pleural effusion (4%, 2/50), pleuritis (4%, 2/50) and cystic adenomatoid malformation (2%, 1/50) by chest radiology. Among

them, one patient (2%, 1/ 50) presented with emphysema in azygos lobe, and 8 children (16%, 8/50) diagnosed as azygos lobe infection, which manifested as patchy opacity (14%) and consolidation (2%). (Fig. 1)

We analyzed the clinical characteristics of children with or without azygos lobe infection, and no significant differences between those two groups of patients were found as for sex, age and complications including other respiratory malformation, recurrent respiratory infection, congenital heart disease. In our study, all the children with azygos lobe infection were cured after reasonable anti-infective treatment.

## Discussion

An azygos lobe is a rare anatomic variant of the lung, which forms during the vein penetrates through the upper lobe of the lung and drags the parietal and visceral pleura with it. The lack of understanding leads to the missed diagnosis of azygos lobe. In our study, the azygos lobes of 20 children were not identified in the first imaging examination.

Most studies report that an azygos lobe is not susceptible to disease. Because of the mesoazygos, the azygos lobe may be isolated from pathological processes developing in the rest of the lung tissue, such as the dissemination of pulmonary tuberculosis and other pathogen infection. However, there are still multiple reports about pathological conditions associated within the azygos lobe. Ndiaye reported that the azygos lobe could lead to atelectasis or bronchiectasis if the fissure was too deep to compress the underlying bronchus draining the azygos lobe. Cases of spontaneous pneumothorax, recurrent hemoptysis and cancer (such as azygos vein aneurysm) associated with an azygos lobe have been reported as well. Pathological processes originating in the azygos lobe, such as carcinoma, may be confined to it.

However, the aforementioned reports were all conducted in adults. To the best of our knowledge, the clinical characteristics about children with azygos lobe has not been reported so far. In our study, 14 (28%) patients were found to have azygos lobe incidentally, while the other 36 cases were diagnosed as pulmonary infections. Furthermore, there were no cases of atelectasis, bronchiectasis, pneumothorax and neoplasm associated with the azygos lobe. And the infection of azygos lobe, manifested as patchy opacity (14%) and consolidation (2%), was found in 8 cases, which was different from most previous literature reports. Among them 3 patients had infection only confined to the azygos lobe, and another 5 patients also had infection of other lung lobes. It was hard to tell whether the infection of the azygos lobe was primary infection or caused by the dissemination of other lung lobe infection. But there were evidences that the azygos lobe also underwent pathological changes, which should not be ignored. To explore risk factors related to the azygos lobe infection, characteristics of those cases with and without azygos lobe infections were compared. However, no significant risk factors were found. The cause of the azygos lobe infection was still unknown.

Interestingly, we found that some cases with the azygos lobe also had other abnormalities including congenital heart disease, Down syndrome, and other respiratory malformations. Whether formation of azygos lobe is related to genetic variation is a question worthy of further exploration.

RRTIs in children remain a great challenge to pediatricians. The causes of RRTIs vary a lot including malnutrition, tobacco exposure, low social-economic status, immunodeficiency, and respiratory malformations are one of the most important causes of RRTIs. In our study, we found 8 patients with azygos lobe had a history of RRTIs. It reminds us that we should consider the existence of azygos lobe when managing children with RRTIs and chest radiology should be conducted in time to confirm the diagnosis.

Azygos lobe does not require special treatment unless it causes significant diseases, such as spontaneous pneumothorax, recurrent infection, cancer, and so on. So far, there are limited reports on the treatment of azygos lobe. Thoracotomy and video-assisted thoracoscopic surgery lobectomy to treat lung cancer originating from azygos lobe were reported. In addition, lobectomy was also recommended in patients with recurrent infection of azygos lobe and spontaneous pneumothorax associated with azygos lobe. In our study, all the children with azygos lobe infection were cured after reasonable anti-infective treatment. However, the long-term prognosis requires further monitoring and study.

## Conclusion

Although rarely associated with pathological conditions, there were still a small number of infective azygos lobe cases in our study. Some of them were found to have other organ malformations as well which needed further monitoring and study. Preoperative awareness of an azygos lobe is very important for physicians and thoracic surgeons.

## Abbreviations

RRTIs  
Recurrent respiratory infections

## Declarations

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### Ethics approval and consents to participate

This study was performed in accordance with the ethical standards of the Declaration of Helsinki and approved by the Institutional Review Board/Ethics Committee affiliated with West China Second

University Hospital, Sichuan University. The informed consents were signed by all the participants.

### **Authors' contributions**

Study conception and design: LW, LC, HL; data collection: LW and FZ; data analysis and interpretation: LW and LC; manuscript drafting: LW; critical manuscript revision: LW, LC, HL; final manuscript approval: all authors.

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### **Availability of data and materials**

The dataset used for the current study is available from the corresponding author on reasonable request.

### **Consent for publication**

Not applicable.

### **Competing interests**

The authors declare that they have no competing interests related to this work.

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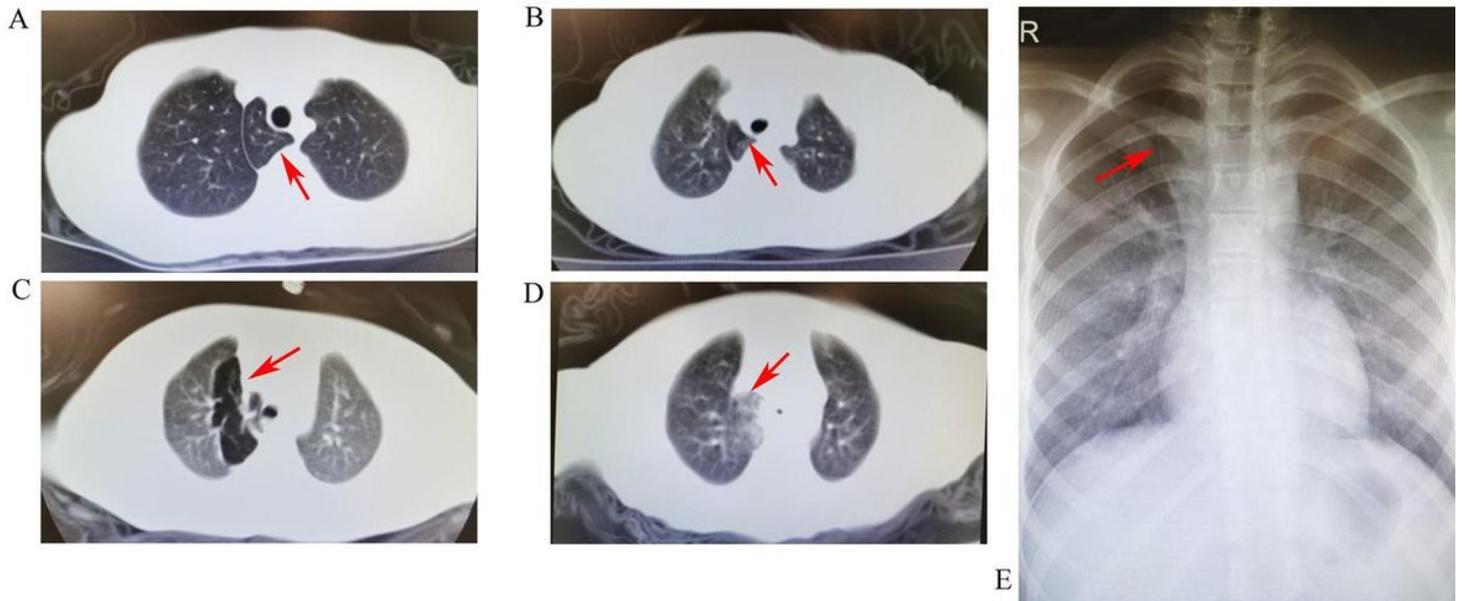
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## Figures



## Figure 1

Computed chest tomography and chest X-ray of showing azygos lobe. A. Computed tomogram of thorax showing azygos lobe in the apicomedial portion of the right lung, separated from rest of the lung by azygous fissure (arrow); B. Computed tomogram of thorax showing azygos lobe co-existed with effusion of other lung lobes; C. Computed tomogram showing the emphysema of azygos lobe; D. Computed tomogram showing the infection of azygos lobe, presented with patchy opacity in azygos lobe; E. Chest X-ray with inverted comma sign / tear drop sign in right upper lobe signifying azygos lobe (arrow).