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A Long-Term Follow-up Study of Childhood Bronchiectasis

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Objective: This study represents the experience of a tertiary care center in Saudi Arabia on non-cystic fibrosis bronchiectasis.

Method: A retrospective review of all patients with confirmed Non-Cystic Fibrosis (Non-CF) bronchiectasis by chest x-ray and or CT chest in a pulmonary clinic for the period 1993-2005 at a tertiary care center in Riyadh.

Result: A total of 151 cases were diagnosed as Non-CF bronchiectasis. Seventy-five (49.7%) were males, 76 (50.3%) were females. Hundred forty-eight (98%) are alive and three (2%) died. The southwestern regions constituted 72 (50%) of the cases. There is a period (5±3.2) years between the start of symptoms and the diagnosis of bronchiectasis. More than two-third of the patients had cough, tachypnea, wheezing, sputum production and failure to thrive. Ninety-one (60%) had associated disease: pulmonary diseases in 48 (32%), immunodefficiency in 27 (18%), CNS in 18 (12%), cardiac in 12 (8%), and asthma in 103 (68%) of the patients. Left lower lobes was commonly involved in 114 (76%). Sixty-eight (67%) was found to have sinusitis. Forty-nine (32%) developed gastroesophgeal reflux (GER). Hemophilus influenza was cultured in 56 (37%); strept pneumoniae in 25 (17%), and pseudomonas aeruginosa in 24 (16%) of the patients. Eighty percent of the patients who had pulmonary function test had abnormal changes. Disease progression was related to development of symptoms before 5 years of age, persistent atelectasis, and right lower lobe involvement ($p < 0.05$).

Conclusion: Non-CF bronchiectasis is a common problem in Saudi Arabia. Early recognition and institution of treatment with proper vaccination of available anti-bacterial and anti-viral vaccines are encouraged to prevent progression of the disease.

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Bronchiectasis was called an orphan disease for the last two decades as its incidence decreased markedly and became an uncommon clinical entity among adults and children in developed countries[1]. It is defined as a permanent dilatation of the bronchi, which typically involves the second to sixth order of segmental bronchi[2]. It was first described by Laënnec in 1819 based on examination of postmortem specimens[3]. Bierrring (1956)

studied 151 patients in Copenhagen following pneumonia and found only one child (0.7 %) to have bronchiectasis[4]. Post infectious bronchiectasis has been reported in many cases[5]. Infection due to pertussis, measles, influenza, Tuberculosis and Adeno virus have been reported to cause bronchiectasis in the early 1950-1960s[5]. Fileld noted a dramatic decrease in admission rates, for bronchiectasis at five British hospitals from an average of 48 per 10,000 in 1952 to 10 per 10,000 total pediatric admissions in 1960[6]. She speculated that improved treatment of lower respiratory tract infections made possible by the increased availability of broad-spectrum antibiotics during that period[6-8]. Other contributing factors include the prevention of measles and pertussis through immunization and the marked decrease in primary pulmonary tuberculosis in the pediatric population which was brought about by better public health measures and improved treatment regimens for this disease[3]. Published reports from some developing countries suggest that childhood bronchiectasis may not be disappearing, and that it represents a more common problem than in developed countries. Karakoc from Turkey described 23 children with bronchiectasis and found that factors other than infections have contributed to the development of bronchiectasis, such as Immunodefficiency, primary ciliary dyskinesia and asthma[2]. A report by Dawson from United Arab Emirates, Abu Dhabi region, described 32 children with bronchiectasis from a population of 300,000[9]. He found that congenital anomalies of the respiratory system, prematurity, Immunodefficiency were some of the factors that contributed to the cause of the disease in addition to viral or bacterial infections[9].

In this study, we present the experience of a tertiary care center in Saudi Arabia on the etiological factors that contributed to childhood bronchiectasis.

METHOD

A retrospective review of charts for all patients referred to the pulmonary clinic for evaluation of recurrent chest infection during the period January 1993 to August 2005 at King Faisal specialist hospital and research center (KFSH&RC). Personal data, radiological patterns, associated diseases, and pulmonary function test data (PFT) were collected.

SPSS program for Windows (release 11.0.0) was used for data analysis. Chi-Square (X2) was used to compare categorical variables. Results were presented at a level of significance of $p = <0.05$.

Progression of disease is a qualitative measurement, defined as a radiological deterioration with more lobes involved in addition to clinical deterioration with increased sputum production, cough and or fever.

PFT severity is a quantitative measurement of airflow in PFT:

Mild lung changes are defined as forced expiratory volume in one second forced expiratory volume in one second (FEV1) as 65-75% of predicted values.

Moderate lung changes are FEV1 = 45-65% have been predicted.

Severe lung changes are FEV1 < 45% have been predicted.

PFT Types are considered normal if forced vital capacity (FVC) and FEV1 = 80-120% predicted values for age and height.
Abnormal PFT: If FVC and FEV1 <75-80% predicted values for age and height

RESULT

A total of 900 cases were referred with recurrent chest infection to pulmonary clinic between January 1993 to August 2005. Two hundred patients were diagnosed to have Cystic fibrosis (CF). One hundred fifty-one cases were diagnosed as non-CF bronchiectasis based on chest x-ray in 151 (100%) and or CT chest in 145 (96%) of the patients. Seventy-five (49.7%) were males, 76 (50.3%) were females. One hundred forty-eight (98%) are alive and three had (2%) died. One hundred forty-four (95%) were Saudi and 7(5%) were non-Saudi. One hundred forty (93%) were full term. Twenty-two (14.6%) from the Eastern region, 26 (17.2%) from the central region, 39 (25.8%) from the western region, 33 (21.9%) from the southern region, and four (2.6%) were from neighboring countries. Ninety-eight (65%) of the families were consanquinous, with 18 patients (12%) had 1-2 siblings with bronchiectasis and five patients had 3-4 siblings with similar disease. Age when symptoms started 2.3 ± 2.2 years. Age at referral to our center was 6.3 ± 4 years. Age of bronchiectasis diagnosis was 7.3 ± 4.1 years. There is a period of (5± 3.2) years between the start of symptoms to the diagnosis of bronchiectasis. Period of follow up 5.5 ± 3.9 years. More than two-third of the patients presented with cough, tachypnea, wheezing, sputum production and failure to thrive. Clubbing was found in 50 (33%) of the patients. Cyanosis and oxygen requirement was reported in 35 (23%) of the patients. Hemoptysis was only reported in seven (5%) of the cases. Ninety-one (60%) had associated diseases. Pulmonary diseases were found in 48 (32%), Immunodefficiency in 27 (18%), Central nervous system (CNS) in 18 (12%), cardiac in 12 (8%), skeletal anomalies in 10 (7%) and asthma in 103 (68%) of the patients (table 1 and 2).

Table (1): Comparisons of disease associations with other developing countries

Associated disease	Banjar 2005 KSA Total (151 patients)	Karakoc 2001 Turkey Total (23)	Dawson 1996 Abu-Dhabi/ UAE Total (32)
Cardiac	12 (8%)		
Pulmonary	48 (32%)	8 (34%)	6 (19%)
Kartagener syndrome	5 (3%)	3 (13%)	
Infections	77 (51%)	8 (34%)	7 (22%0
CNS	18 (12%)		
FBA	6 (4%)		
Immunodefficiency	27 (18%)		3(9%)
Skeletal	10 (7%)		
Asthma	103 (68%)	4 (17%)	
Other	78 (52%)		

KSA- Kingdom of Saudi Arabia
UAE- United Arab Emirates
CNS- Central nervous system
FBA- Foreign body aspiration

Table (2): Bronchiectasis and disease association (Total 91 (60%) Patients)

Disease association	Number	Disease association	Number
Pulmonary:		**Immunodefficiency:**	
Kartagener	4	Hypogammaglobulinemia	3
FBA	6	SCIDS	3
Immotile cilia syndrome	17	HIV	1
Lipid pneumonia	7	Hyper IgE	1
Interstitial pneumonia	2	IgG subclass defficiency	6
ABPA	2	Hyper IgM	2
T.B	2	Whiscott Aldrich syndrome	1
RML syndrome	1	Poor antibodies response	4
TEF repair	4	Common variable-Hypogammaglobulinemia	3
Bronchogenic cyst	2	T-cell deficiency	3
Cystic lung disease	5	Barre lymphocyte syndrome	1
Lung collapse	3		
Prematurity	3		
Cardiac Diseases:		**Central nervous system disease:**	
Dextrocardia	4	Cerebral palsy/ seizure disorder	4
Congestive heart failure	1	Apnea	1
Ventricular septal defect	2	Craniosynostosis	1
Atrial septal defect	1	Cutis laxa/ developmental delay	1
Pulmonary hypertension	1	Down syndrome/ Seizure	2
Mitrale valve prolapse	1	Fatty acid oxidation defect	1
Skeletal:		**Other disease associations:**	
Pectus excavatum	2	Neuroblastoma	1
Scoliosis	4	Antithrombin III defficiency	2
Absent ribs	3	Corrosive ingestion	2
Marfan's syndrome	1	Liver cirrhosis	1
		Ethmoid mucocele	1
		Bullous skin lesion/ septicemia	1

FBA- Foreign body aspiration
ABPA- Allergic Bronchopulmonary Aspergillosis
TB- Tuberculosis
RML- Right middle lobe
TEF- Tracheoesophageal fistula
SCIDS- severe combined Immunodefficiency

Radiological changes reported are : consolidation of one or two lobes in 137 (91%) of the patients, hyperinflation in 103 (68%), interstitial pattern in 49 (33%), atelectasis in 117 (78%), Peribronchial wall thickening in 115 (76%) and lymph node enlargement of the para tracheal region in 33 (22%) of the patients. Left lower lobes (LLL) was commonly involved in 114 (76%), right middle lobe (RML) in 82 (54%), and right lower lobe (RLL) in 76 (50%), lingula in 73 (48%), right upper lobe (RUL) in 39 (26%), and left upper lobe (LUL) in 27 (18%) of the patients. Bilateral lobar involvement in 112 (71%). A total of 102 patients had sinus x-ray, and 18 (12%) had CT sinuses. Sixty-eight (67%) of 102 patients who had sinus x-ray found to have sinusitis. Sixty-nine (46%) of the patients had investigation for gastroesophgeal reflux (GER) by barium swallow and by milk scan in 27 (18%) of the patients. GER was found in 49 (32%) patients, 43 on barium swallow group

and 13 on milk scan group. Twenty-two of 49 patients with GER (45%) required Nissen fundo-plication. Sputum or nasopharyngeal aspiration (less than 4 years) cultures were done on 105 (70%). Hemophilus influenza (H-flue) was cultured in 56 (37%), Streptococcus pneumoniae in 25 (17%), Pseudomonas aeruginosa in 24 (16%), Branhamella Cattarrhales in 13 (9%), Staphylococcus aureus (Staph.) in 11 (7%) and Methicillin resistant staphylococcus aureus (MRSA) in three (2%) patients. Candida albicans in two (1%) of the patients. Viral cultures were done in 33 (22%) of the patients: Respiratory Syncytial virus in three (9%), and Enterovirus in one (3%). Disease progression developed in 72 (48%) of the patients and it was related to development of symptoms before five years of age, persistent atelectasis of the affected lobes, and involvement of RLL with bronchiectasis ($p < 0.05$). Unilateral Lobectomy was done in 21 (14%) patients whereas bilateral lobectomies in three (2%). Recurrent otitis media was reported in 12 (8%) patients. Blood gas at presentation showed a picture of mild compensated respiratory acidosis with metabolic alkalosis, Mean values (±SD) are: PH= 7.38 (±0.06), PaCO2 = 6.54 (± 2), PaO2 of 9.81 (±3.95), HCO3 of 27.93 (± 5.65), BE of 2.46 (± 4.57), and O2 saturation of 90 (±17.3).

PFT changes

Seventy-seven (49%) of the patients were able to do pulmonary function test (PFT). Sixty-eight (88%) of them had abnormal PFT changes (table 3).

Table (3): PFT values at presentation (Total 77 patients)

Variable	Mean (SD)	Median	Minimum	Maximum
FVC	66.7 (18.5)	67	28	114
FEV	64.8 (20.3)	66	33	124
FEV1 / FVC	98 (15.6)	100	41	121
MMEF 25-75%	53 (27.1)	52	9	118
PEF	66.5 (20.3)	67	13	112
% CHANGE VENT in FEV1	15.6 (22.9)	9.5	-31	98
% CHANGE VENT in MMEF 25-75%	24.2 (23.9)	24	-10	65
FRC	106.3 (20.7)	107	64	156
RV	151.5 (40.1)	144.5	80	237
TLC	84.5 (11.6)	84.5	53	109
RV/ TLC ratio	46.4 (12.1)	44.5	27	77
RV/ TLC (%)	179.9 (44.9)	170.5	101	288

FVC- Forced vital capacity
FEV1- Forced expiratory volume in one second
MMEF- Maximum mid expiratory flow
PEF- Peak expiratory flow
% Ventolin- percentage of change in FEV1 values after administration of Ventolin
FRC- Functional residual capacity
RV-Residual volume
TLC- Total lung capacity
RV/TLC- The ratio of RV/TLC in percentage and actual values

Seventeen (22%) had obstructive lung changes, 14 (18%) had restrictive lung changes, and 37 (48%) had combined obstructive and restrictive lung changes. Sixteen (21%) had mild PFT changes, 30 (39%) moderate lung changes and 22 (28.5%) had severe lung changes. There was a decrease in all PFT parameters by 1-2% at follow up (table 4).

Table (4): PFT values at follow up
Total of 55 patients

Variable	Mean (SD)	Median	Minimum	Maximum
PFT	1.6 (.5)	2	1	2
FVC	64 (19)	66	28	111
FEV1	60.9 (19.7)	61	26	106
FEV1 / FVC	95.4 (14)	98	54	118
MMEF25-75%	42 (25.7)	35	7	123
PEF	64.4 (21.4)	66	10	105
% CHANGE VENT in FEV1	22 (27.5)	13.5	-11	98
% CHANGE VENT in MMEF 25-75%	25.9 (32.9)	17.5	-25	83
FRC	106.7 (31.7)	102	62	212
RV	153.2 (61.9)	141	81	400
TLC	83.3 (15.1)	83	52	134
RV/ TLC ratio	45.9 (11.7)	42.5	26	79
RV/ TLC (%)	181.2 (44.8)	171	108	302

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RV/TLC- The ratio of RV/TLC in percentage and actual values

Male patients have shown more severe PFT changes compared to females ($p < 0.05$) (table 5).

Table (5): Correlation of PFT severity to different factors (Total 77 patients)

Factors	Mild changes	Moderate/ severe	Total	P values
Male Female Total	8 (21%) 16 (41%) 24 (31%)	30 (79%) 23 (59%) 53 (69%)	38 (100%) 39 (100%) 77 (100%)	0.05
Sinusitis	14 (32%)	30 (68%)	44 (100%)	0.8
GER	9 (38%)	15 (62%)	24 (100%)	0.4
Ventilation	3 (43%)	4 (57%)	7 (100%)	0.4
Disease association	9 (24%)	28 (76%)	37 (100%)	0.2
Asthma	18 (32%)	39 (68%)	57 (100%)	0.8
Required lobectomy	3 (18%)	14 (82%)	17 (100%)	0.1
O_2 requirement	1 (14%)	6 (86%)	7 (100%)	0.3
Consanguinity	12 (25%)	36 (75%)	48 (100%)	0.1

GER- Gastroesophageal reflux
Mild PFT changes- FEV1 65-75% of predicted values for age and height
Moderate/severe PFT changes- FEV1 35-65% of predicted values for age and height

Patients with positive respiratory culture for H-flue had more abnormal PFT changes compared to those who had negative culture for this type of bacteria at presentation but not at follow up and it was not related to their PFT severity (table 6).

Table (6): Correlation of Hemophilus influenza and PFT
Total 77 patients

PFT	Type and severity	Number (%)	P value
Severity at presentation	Mild	11 (27%)	
	Mod / Severe	30 (73%)	0.2
Severity at follow up	Mild	11 (36%)	
	Mod / Severe	20 (64%)	0.3
PFT Type at presentation	Normal	2 (5%)	
	Abnormal	39 (95%)	0.04
PFT Type at follow up	Normal	2 (6%)	
	Abnormal	30 (94%)	0.7

Mild PFT changes- FEV1 65-75% of predicted values for age and height
Moderate / severe PFT changes- FEV1 35-65% of predicted values for age and height
Normal PFT Type: If FVC and FEV1 = 80-120% predicted values for age and height.
Abnormal PFT Type: If FVC and FEV1 <75-80% predicted values for age and height

Involvement of the lingula shared the least factors to severity of PFT changes ($p < 0.03$) (table 7).

Table (7): Correlation of lobes involved and severity of PFT (Total 77 patients)

Lobes involved	Mild changes	Moderate/ severe	Total	P value
At presentation				
RUL	4 (25%)	12 (75%)	16 (100%)	0.5
RML	16 (33%)	32 (67%)	48 (100%)	0.5
RLL	12 (30%)	28 (70%)	40 (100%)	0.9
LUL	4 (24%)	13 (76%)	17 (100%)	0.4
Lingula	9 (24%)	28 (76%)	37 (100%)	0.1
LLL	19 (33%)	38 (67%)	57 (100%)	0.4
At follow-up				
RUL	4 (29%)	10 (71%)	14 (100%)	0.9
RML	10 (27%)	27 (73%)	37 (100%)	0.6
RLL	8 (30%)	19 (70%)	27 (100%)	0.9
LUL	2 (15%)	11 (85%)	13 (100%)	0.2
Lingula	4 (15%)	22 (85%)	26 (100%)	0.03
LLL	11 (26%)	31 (74%)	42 (100%)	0.3

RUL: Right upper lobe
RML: Right middle lobe
RLL: Right lower lobe
LUL: Left upper lobe
LLL: Left lower lobe

DISCUSSION

In our study, bronchiectasis incidence was found to be one in four cases that presented with recurrent chest infection to our center, which makes it a common problem in this part of the world. Bacterial infection with the common respiratory organisms such as Staph aureus, H-flue, Pneumococcus, and Pseudomonas were found to be common bacteria cultured from 51% of the patients. The Southwestern region accounted for 50% of the reported cases. Environmental factors such as humidity and crowdness during Pilgrimage time may have contributed to such increase in its incidence. Recurrent aspiration pneumonia due CNS anomalies or seizure is described for the first time in the literature and might be related to recurrent aspiration of secretions due to swallowing incoordination and or GER. This study agrees with other studies of early symptoms before five years of age in 83% of our population with a delay of diagnosis of bronchiectasis by an average of 5-10 years[1,2,8]**.** In one study among 46 Alaskan children reported with bronchiectasis, each child experienced an average of nine lower respiratory illness before the diagnosis of bronchiectasis was made and their chest radiographs suggested that the lobes were abnormal at two years of age were more likely to become Bronchiectatic than a lobe involved during any single infection. Presumably multiple bacterial and or viral infections lead to cumulative airway injury, narrowing and poor mucous clearance, setting the stage for evolution of bronchiectasis several years late[1,8].

Most of the patients had bilateral lobar involvement and severe PFT changes at presentation. Fifty percent of our patients had radiological and clinical progression in spite of medical treatment with antibiotic, which may suggest the value of surgical intervention in patients with progressive disease, lobectomy has been done in only 16% of our patients compared to 60-70% in other studies[2]. Asthma was common association in 68% of the patients, which is in accordance with other studies and treatment with inhaled steroid, and B2 agonist may need to be considered in some patients[10-12]. Immunodefficiency is common in our country due to consanguinity and found to be the second most common disease association after pulmonary disease.

Sinusitis was also a common presentation (68%) and such patients may need to be treated for a longer period of time as suggested by other studies for 4-6 week[3]. Persistent atelectasis of the affected lobe has been contributing to the development of bronchiectasis in our patients, which may warrant encouragement of chest physiotherapy, and postural drainage in patients with such a problem. Atelectasis is commonly found in many patients with pneumonia, aspiration or asthma and repeat chest x-ray should be done after clinical improvement to ensure the re-expansion of the atelectatic part of the lung. Gastroesophageal reflux and recurrent aspiration is found in 32% of our patients and may have contributed to the development of bronchiectasis or complicated its progression. More than two-third of the patients presented with abnormal PFT and moderate to severe changes.

H-flue played an important role in the development of abnormal PFT at presentation but not to the progression of disease.

CONCLUSION

Non-CF bronchiectasis is a common problem in Saudi Arabia. Early recognition and institution of treatment with proper vaccination of available anti-bacterial and anti-viral vaccines are encouraged to prevent progression of the disease.

A case control study needs to be done to identify the actual risk factors of developing such disease in our country and efforts should be made to early diagnosis, awareness of contributing factors and early treatment or referral before development of progression.

REFERENCES

1. Callahan CW, Redding G. Bronchiectasis in children: Orphan disease or persistent problem? Pediatr Pulmonol 2002;33:492-6.
2. Karakoc GB, Yilmaz M, Altintas DU, et al. Bronchiectasis: Still a problem. Pediatr Pulmonol 2001;32: 175-8.
3. Brown MA, Leman RJ. Bronchiectasis. Chernick V, Boat T, eds. In: Kendig's disorder of the respiratory tract in children, 6th ed, Philadelphia: WB Saunders; 1998; 538-60.
4. Biering A. Childhood pneumonia, including pertussis, pneumonia and bronchiectasis: a follow-up study of 151 patients. Acta Pediatr 1956; 45:348-51.

5. Barker AF, Bardana EJ. State of the art, Bronchiectasis: Update of an Orphan disease. Am Rev Respir Dis 1988; 137: 969-78.
6. Field CE. Bronchiectasis: Third report on a follow-up study of medical and surgical cases from childhood. Arch Dis Child 1969;44: 551-5.
7. Clark NS. Bronchiectasis in childhood. Br med J 1963; 1: 80-7.
8. Singleton R, Morris A, Redding G, et al. Bronchiectasis in Alaska Native children: causes and clinical courses. Pediatr Pulmonol 2000;29:182-7.
9. Dawson KP, Bakalinova D. Child bronchiectasis in a desert location. Middle East pediatrics 1996;1:6-8.
10. Ip MSM, So SY, Lam WK, et al. High prevalence of asthma in patients with bronchiectasis in Hong Kong. Eur Respir J. 1992;5: 418-23.
11. Bahous J, Cartier A, Pineau L, et al. Pulmonary function test and airway responsiveness to methacholine in chronic bronchiectasis of the adults. Bull Eur Physiopath Respir 1984;20:375-80.
12. Varpela E, Laitinen LA, Leakinen H, et al. Asthma, allergy and bronchial hyper-reactivity in bronchiectasis: a controlled study. Thorax 1989; 44: 948-51.

Table (1): Comparisons of disease associations with other developing countries

Associated disease	Banjar 2005 KSA Total (151 patients)	Karakoc 2001 Turkey Total (23)	Dawson 1996 Abu-Dhabi/ UAE Total (32)
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Kartagener syndrome	5 (3%)	3 (13%)	
Infections	77 (51%)	8 (34%)	7 (22%0
CNS	18 (12%)		
FBA	6 (4%)		
Immunodefficiency	27 (18%)		3(9%)
Skeletal	10 (7%)		
Asthma	103 (68%)	4 (17%)	
Other	78 (52%)		

KSA- Kingdom of Saudi Arabia
UAE- United Arab Emirates
CNS- Central nervous system
FBA- Foreign body aspiration

Table (2): Bronchiectasis and disease association (Total 91 (60%) Patients)

Disease association	Number	Disease association	Number
Pulmonary:		**Immunodefficiency:**	
Kartagener	4	Hypogammaglobulinemia	3
FBA	6	SCIDS	3
Immotile cilia syndrome	17	HIV	1
Lipid pneumonia	7	Hyper IgE	1
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Bronchogenic cyst	2	T-cell deficiency	3
Cystic lung disease	5	Barre lymphocyte syndrome	1
Lung collapse	3		
Prematurity	3		
Cardiac Diseases:		**Central nervous system disease:**	
Dextrocardia	4	Cerebral palsy/ seizure disorder	4
Congestive heart failure	1	Apnea	1
Ventricular septal defect	2	Craniosynostosis	1
Atrial septal defect	1	Cutis laxa/ developmental delay	1
Pulmonary hypertension	1	Down syndrome/ Seizure	2
Mitrale valve prolapse	1	Fatty acid oxidation defect	1
Skeletal:		**Other disease associations:**	
Pectus excavatum	2	Neuroblastoma	1
Scoliosis	4	Antithrombin III defficiency	2
Absent ribs	3	Corrosive ingestion	2
Marfan's syndrome	1	Liver cirrhosis	1
		Ethmoid mucocele	1
		Bullous skin lesion/ septicemia	1

FBA- Foreign body aspiration
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Table (3): PFT values at presentation (Total 77 patients)

Variable	Mean (SD)	Median	Minimum	Maximum
FVC	66.7 (18.5)	67	28	114
FEV	64.8 (20.3)	66	33	124
FEV1 / FVC	98 (15.6)	100	41	121
MMEF 25-75%	53 (27.1)	52	9	118
PEF	66.5 (20.3)	67	13	112
% CHANGE VENT in FEV1	15.6 (22.9)	9.5	-31	98
% CHANGE VENT in MMEF 25-75%	24.2 (23.9)	24	-10	65
FRC	106.3 (20.7)	107	64	156
RV	151.5 (40.1)	144.5	80	237
TLC	84.5 (11.6)	84.5	53	109
RV/ TLC ratio	46.4 (12.1)	44.5	27	77
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Table (4): PFT values at follow up
Total of 55 patients

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PFT	1.6 (.5)	2	1	2
FVC	64 (19)	66	28	111
FEV1	60.9 (19.7)	61	26	106
FEV1 / FVC	95.4 (14)	98	54	118
$MMEF_{25-75\%}$	42 (25.7)	35	7	123
PEF	64.4 (21.4)	66	10	105
% CHANGE VENT in FEV1	22 (27.5)	13.5	-11	98
% CHANGE VENT in $MMEF_{25-75\%}$	25.9 (32.9)	17.5	-25	83
FRC	106.7 (31.7)	102	62	212
RV	153.2 (61.9)	141	81	400
TLC	83.3 (15.1)	83	52	134
RV/ TLC ratio	45.9 (11.7)	42.5	26	79
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Factors	Mild changes	Moderate/ severe	Total	P values
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Sinusitis	14 (32%)	30 (68%)	44 (100%)	0.8
GER	9 (38%)	15 (62%)	24 (100%)	0.4
Ventilation	3 (43%)	4 (57%)	7 (100%)	0.4
Disease association	9 (24%)	28 (76%)	37 (100%)	0.2
Asthma	18 (32%)	39 (68%)	57 (100%)	0.8
Required lobectomy	3 (18%)	14 (82%)	17 (100%)	0.1
O_2 requirement	1 (14%)	6 (86%)	7 (100%)	0.3
Consanguinity	12 (25%)	36 (75%)	48 (100%)	0.1

GER- Gastroesophageal reflux
Mild PFT changes- FEV1 65-75% of predicted values for age and height
Moderate/severe PFT changes- FEV1 35-65% of predicted values for age and height

Table (6): Correlation of Hemophilus influenza and PFT
Total 77 patients

PFT	Type and severity	Number (%)	P value
Severity at presentation	Mild	11 (27%)	
	Mod / Severe	30 (73%)	0.2
Severity at follow up	Mild	11 (36%)	
	Mod / Severe	20 (64%)	0.3
PFT Type at presentation	Normal	2 (5%)	
	Abnormal	39 (95%)	0.04
PFT Type at follow up	Normal	2 (6%)	
	Abnormal	30 (94%)	0.7

Mild PFT changes- FEV1 65-75% of predicted values for age and height
Moderate / severe PFT changes- FEV1 35-65% of predicted values for age and height
Normal PFT Type: If FVC and FEV1 = 80-120% predicted values for age and height.
Abnormal PFT Type: If FVC and FEV1 <75-80% predicted values for age and height

Table (7): Correlation of lobes involved and severity of PFT (Total 77 patients)

Lobes involved	Mild changes	Moderate/ severe	Total	P value
At presentation				
RUL	4 (25%)	12 (75%)	16 (100%)	0.5
RML	16 (33%)	32 (67%)	48 (100%)	0.5
RLL	12 (30%)	28 (70%)	40 (100%)	0.9
LUL	4 (24%)	13 (76%)	17 (100%)	0.4
Lingula	9 (24%)	28 (76%)	37 (100%)	0.1
LLL	19 (33%)	38 (67%)	57 (100%)	0.4
At follow-up				
RUL	4 (29%)	10 (71%)	14 (100%)	0.9
RML	10 (27%)	27 (73%)	37 (100%)	0.6
RLL	8 (30%)	19 (70%)	27 (100%)	0.9
LUL	2 (15%)	11 (85%)	13 (100%)	0.2
Lingula	4 (15%)	22 (85%)	26 (100%)	0.03
LLL	11 (26%)	31 (74%)	42 (100%)	0.3

RUL: Right upper lobe
RML: Right middle lobe
RLL: Right lower lobe
LUL: Left upper lobe
LLL: Left lower lobe