

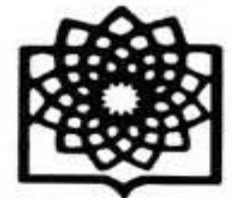
A survey of prevalence and diversity of pulmonary cystic lesions in a referral children's hospital between the years 2004 and 2015

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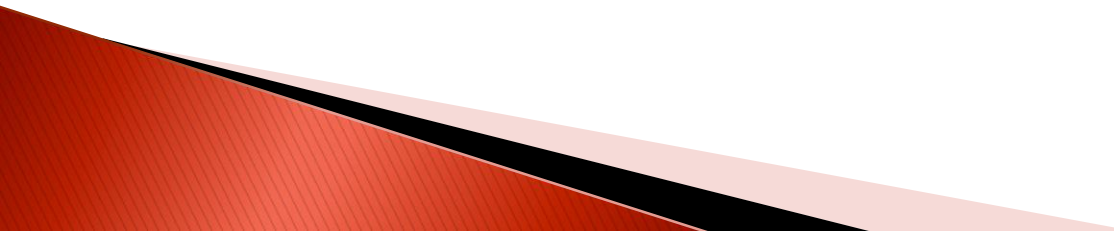
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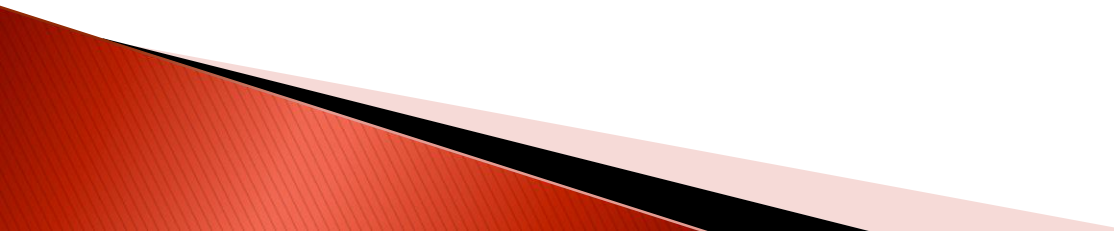




Background

- ▶ Congenital pulmonary lesions
 - ▶ Pulmonary cystic lesions
 - ▶ Origin, nomenclature, pathology and therapeutic approaches
 - ▶ Site of involvement and co-morbidities
 - ▶ Diagnostic imaging tools
 - ▶ Time and way of manifestation
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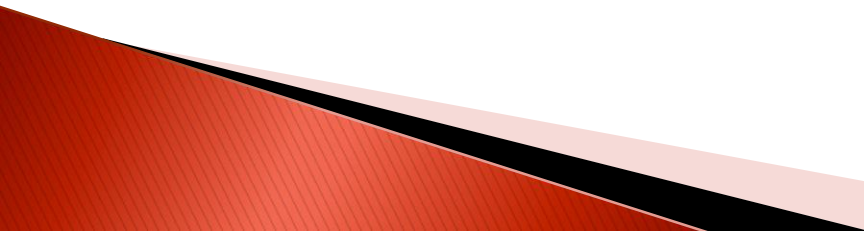
Objectives

- ▶ Prevalence determination
 - ▶ Diagnostic modality application
 - ▶ Diversity and time of presentation
 - ▶ Lesions' specification
 - ▶ Medical team negligence accentuation
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Materials/Methods

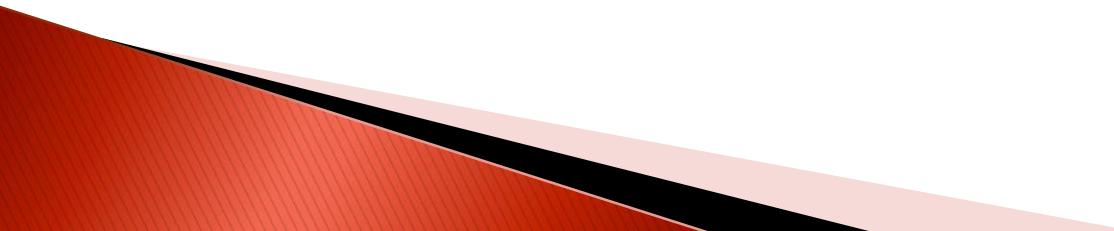
- ▶ All diagnosed or referred patients to Mofid Children's Hospital of Tehran between the years of 2004 and 2015
- ▶ Data pertaining to all patients presenting with congenital lung mass during the 11-year period (2004-2015) was extracted
- ▶ Surgeries were performed by board-certified pediatric surgeons, via transthoracic approach.
- ▶ Imaging and pathology studies were all performed at Mofid Children's hospital
- ▶ Extracted data were; sex, gestational age, age at admission, patient first presentation, length of hospitalization and accompanying features.

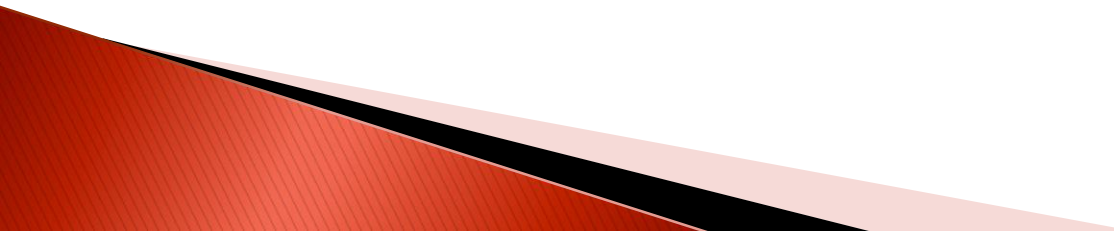
Results

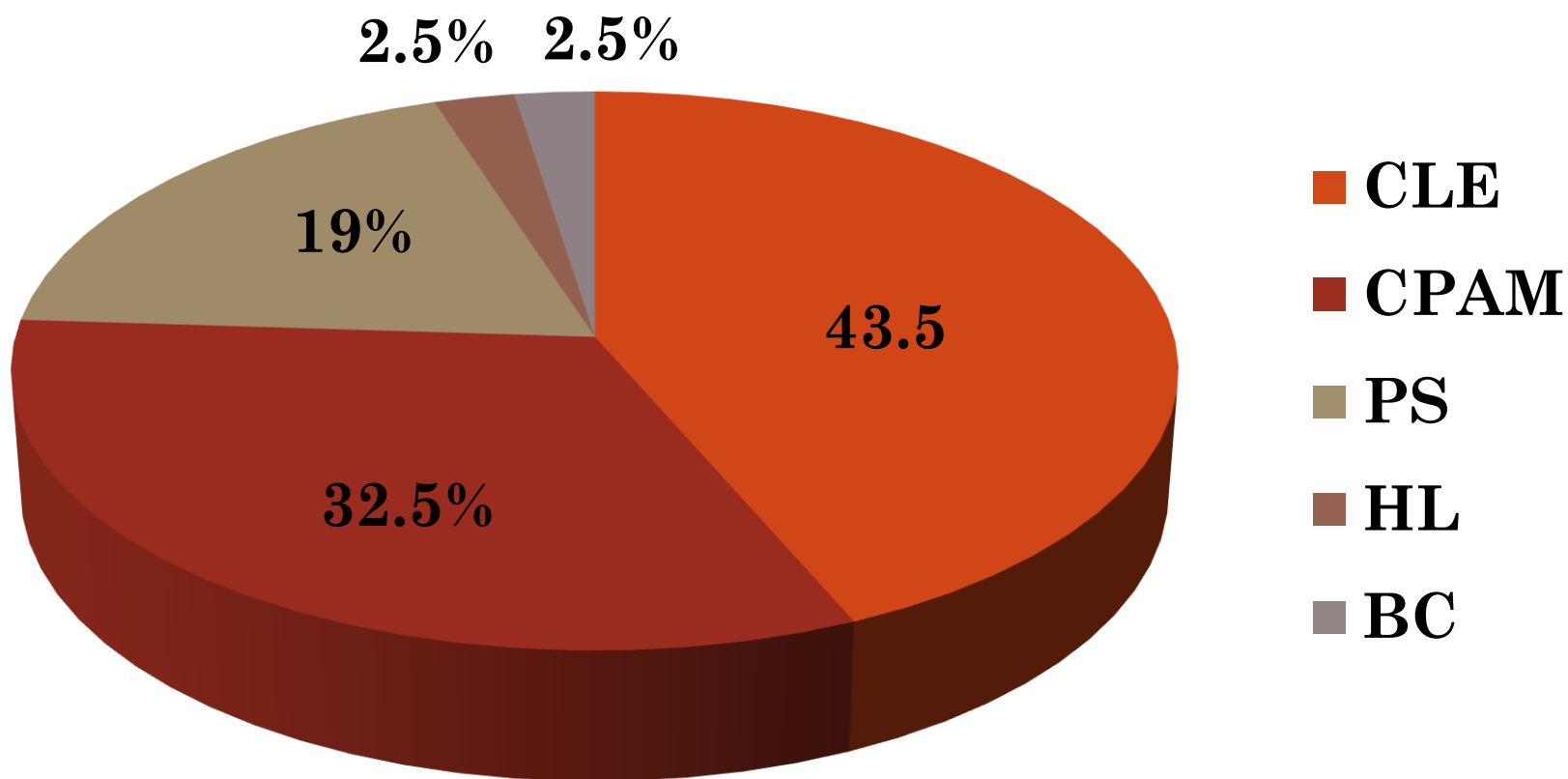
- ▶ Congenital lung mass : **Forty seven** cases
 - ▶ Cystic lesions consisted **78.5 % (37 cases)**
 - ▶ Male predominance (**3 times**)
 - ▶ Lesions were more located in **left** hemithorax (**57% versus 43%**)
 - ▶ The majority manifested with **respiratory complaints**
 - ▶ Age groups of **neonates and infants** constituted almost **equally** and **89%** of all
- 

- ▶ The remaining **11%(Four cases)** were children ;
- ▶ A 3.5 year old female with febrile seizure and accidental finding of **Congenital Lobar Emphysema of left upper lobe** in CXR.
- ▶ Two 5 year old females with recurrent pneumonia who were both diagnosed as **Pulmonary Sequestration of right lower lobe**,
- ▶ An 8 year old female with prolonged fever who was again diagnosed as **Pulmonary Sequestration of left lower lobe**
- ▶ Approximately **three/forth** missed ante-natal diagnosis (most of which found to be CLE *)
- ▶ Only **one/forth** of cases found to have co-morbidities

Pulmonary cystic lesions are divided into :


- ▶ **CLE**(Congenital Lobar Emphysema)
 - ▶ **CPAM**(Congenital Pulmonary Airway Malformation)
 - ▶ **PS**(Pulmonary Sequestration)
 - ▶ **HL**(Hybrid Lesion) : CPAM+PS
 - ▶ **BC**(Bronchogenic Cyst)
- 

- ▶ Distribution frequency of each cystic lesion was as follows :
 - ▶ Congenital Lobar Emphysema(**CLE**),**16 cases**
 - ▶ Congenital Pulmonary Airway Malformation(**CPAM**),**12 cases**
 - ▶ Pulmonary Sequestration(**PS**),**7 cases**
 - ▶ Hybrid Lesions(**HL**),**1 case**
 - ▶ Bronchogenic Cyst(**BC**),**1 case**
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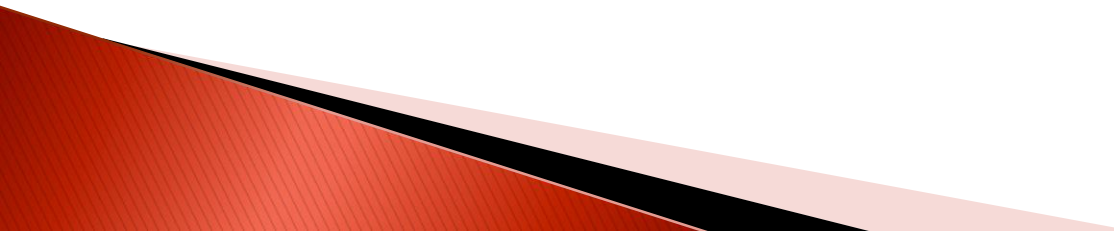


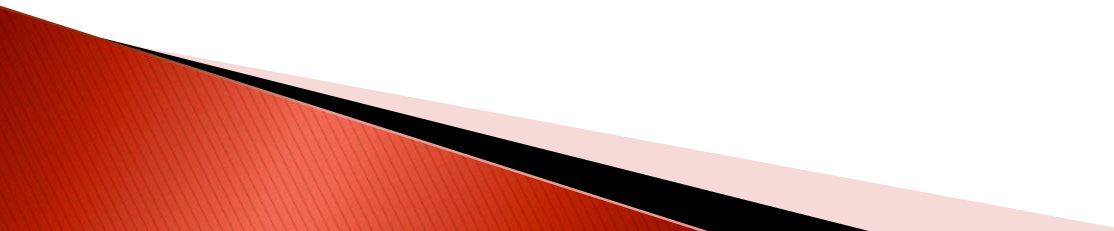
Distribution and Character of Cystic lesions in the survey

Lesion	No.	Right	Left	Bilateral	Age range	M:F	Prenatal diagnosis +/-
CPAM	12	5	6	1	1d-124 d	10:2	4/8
PS	7	4	3	0	19d-8y	4:3	3/4
CLE	16	5	11	0	6d-3.5y	13:3	1/15
HL	1	0	1	0	3d	0:1	1
BC	1	1	0	0	6d	1:0	1
SUM	37	15	21	1	1d-8y	28:9	10/27

- ▶ Unfortunately, **73%** of cases missed to have the opportunity of early diagnosis
 - ▶ Again, **73 %** missed to have pathologic report of their lesions
 - ▶ Accompanying features were Seen in **9 cases(24%)**
 - ▶ **Three** cases of cardiovascular co-morbidities
 - ▶ **Two** cases of hematologic co-morbidities
 - ▶ **Two** cases with CNS involvement
 - ▶ **One** case of hypospadiasis
 - ▶ Co-morbidities were mostly seen with **PS**
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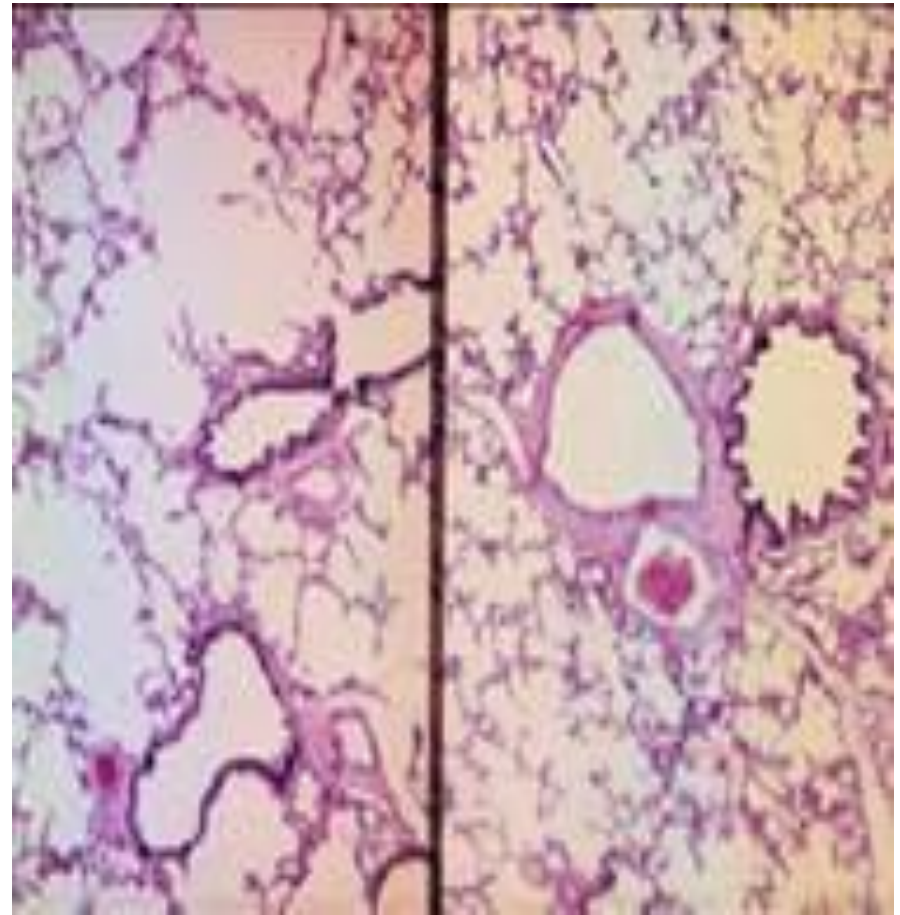
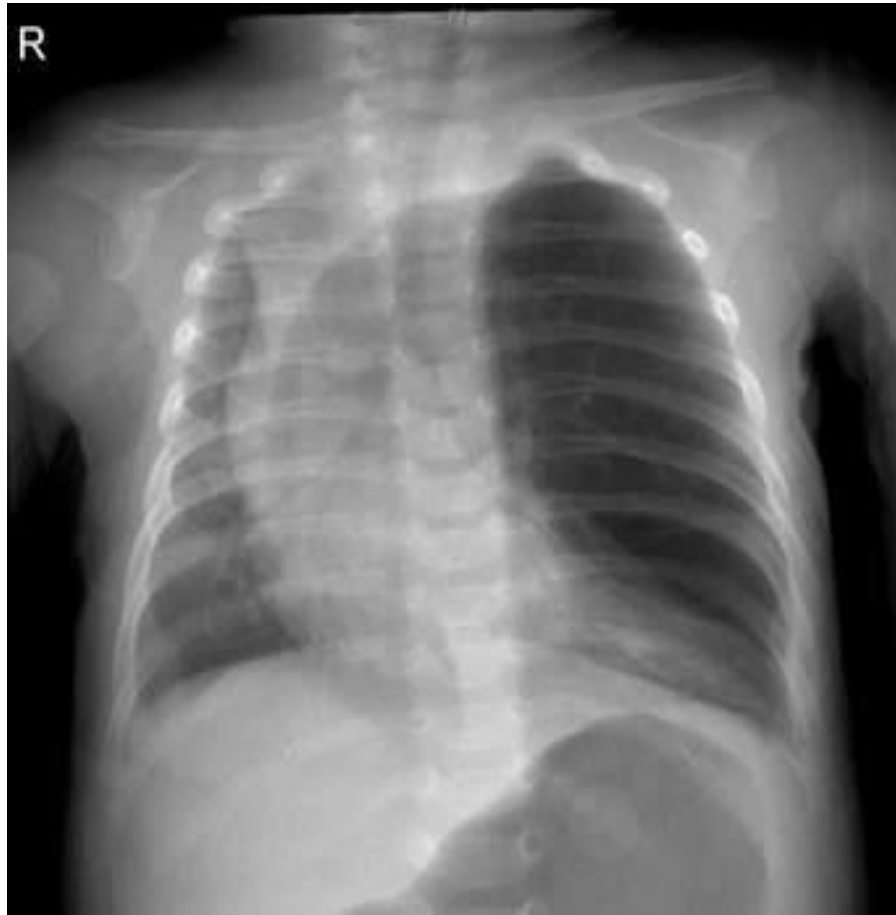
Discussion

- ▶ In a 11-year similar study of a referral center in **Texas**, 42 cases were diagnosed as cystic lesions and the frequencies were as follows:
 - ▶ **PS** (40%)
 - ▶ **CLE** (24%)
 - ▶ **CPAM**(21%)
 - ▶ **BC**(15%)
- 


- ▶ In the study of **Longston**, Male predominance was apparent in all the masses **except for CLE** in which the sexes had the same frequency, while in ours male/female ratio of **CLE** cases was **13/1**.
 - ▶ In another 10 year study in **Saudi Arabia**, of 57 cases, 65% was diagnosed as CLE, 12 % as CCAM, 14% as BC and 9% found to be PS.
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CLE

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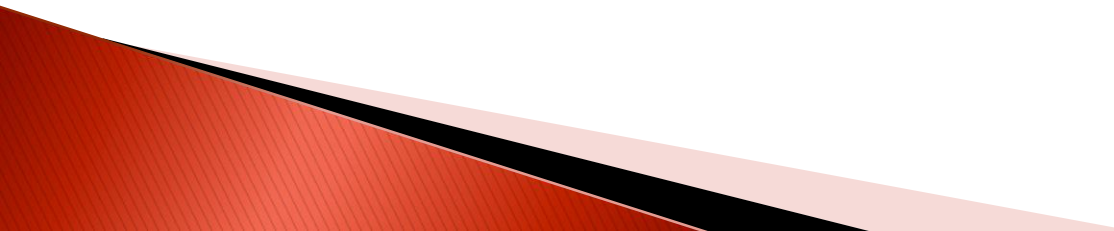


- ▶ Mostly seen in male cases
- ▶ Affected lobes in sequence of prevalence:
 - Left upper lobe (42%)
 - Right middle lobe (35%)
 - Right upper lobe (21%)
 - Lower lobes (2%)
- ▶ Spontaneous regression in the third trimester is not unusual.
- ▶ *Early Xrays may not show hyperlucency but increased opacity due to delayed fluid clearance*

- ▶ May be followed conservatively (Asymptomatic Lesions or the ones with minimal symptoms), on the contrary of CPAMs and PSs with risks of later infection and malignancy transformation
 - ▶ Rarely documented in prenatal sonographies
 - ▶ In **20%** of cases congenital heart diseases accompany
 - ▶ *The differential diagnosis is tension pneumothorax in which the hemodynamic status is also unstable and pulmonary markings are absent*
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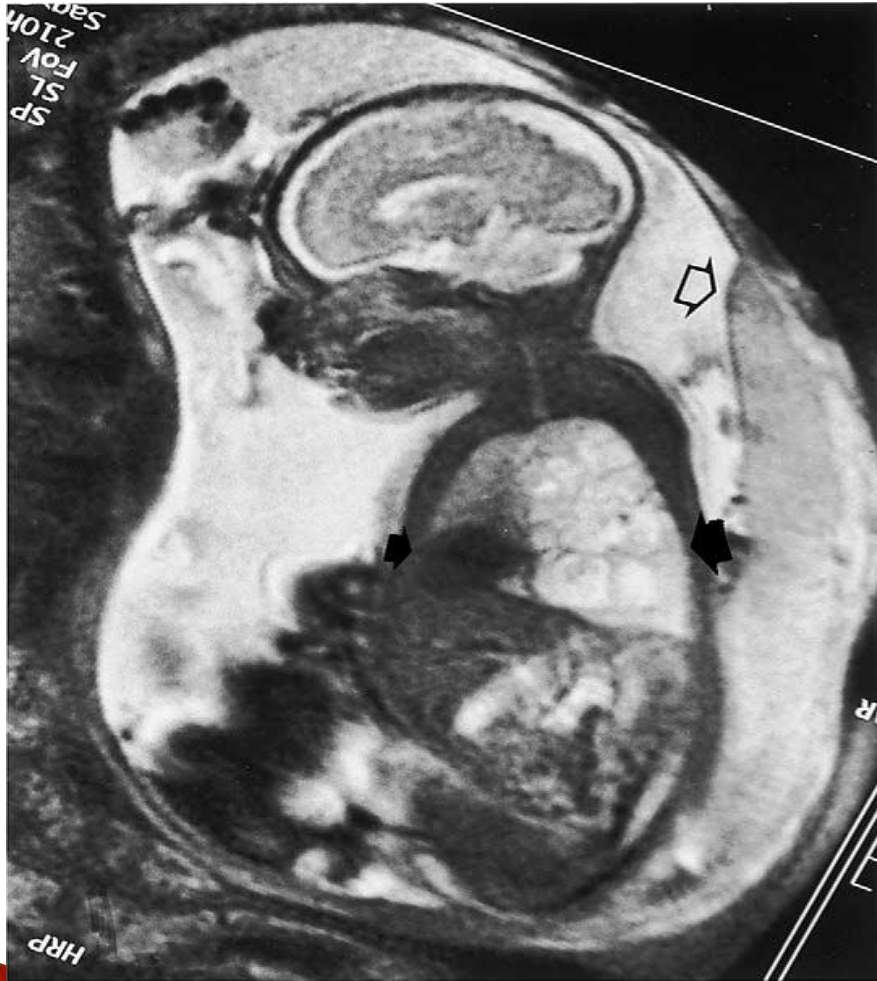
CLE

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- ▶ In our investigation, among **16** cases of CLE,
 - ▶ Only **one** case had prenatal diagnosis,
 - ▶ **13** cases were males,
 - ▶ **11** cases located in Left upper lobe
 - ▶ In this study **none** of the CLE cases were diagnosed to have CHD(Congenital Heart disease)
 - ▶ *Of interest*, was the accompaniment of G6PD deficiency and thrombocytopenia in **two** CLE cases
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CPAM

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A

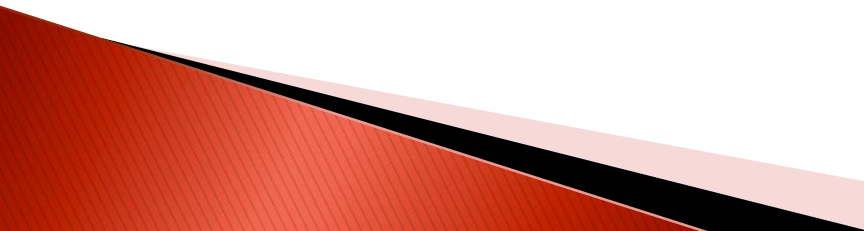


B



CPAM

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- ▶ Incidence of 1/11000 to 1/35000
 - ▶ Previous nomenclature: CCAM
 - ▶ Is subdivided into **5 types**
 - ▶ Types 1,2,3 :Adenomatoid; Types 0,4:Cystic
 - ▶ Also Classified as **microcystic** and **macrocystic** based on gross anatomy and prenatal sonography
 - ▶ Microcystic lesions' size is considered 5 mm and below and have worse prognosis
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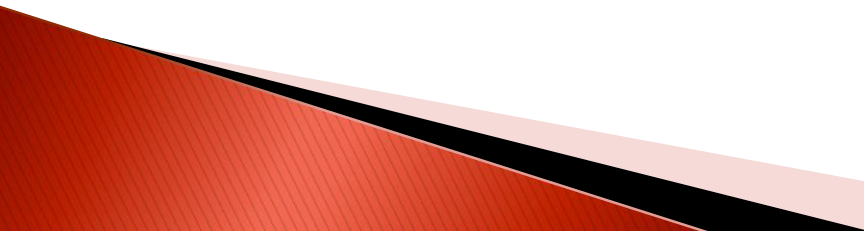
CPAM

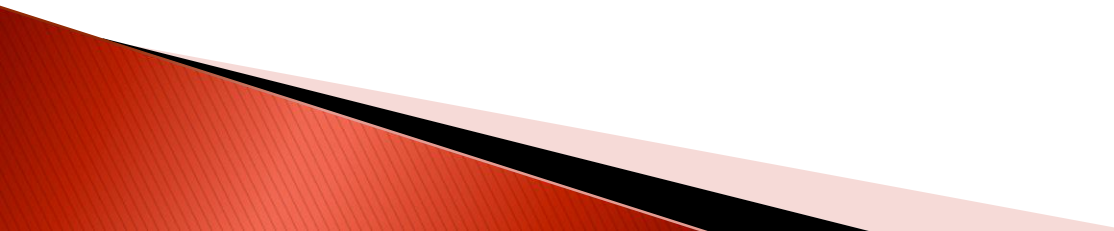
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- ▶ Involves **both** lungs equally, mostly lower lobes
- ▶ Attached to trache-bronchial tree
- ▶ CPAM can regress prenatally(**15%**), stay the same size, or grow and produce hydrops, pulmonary hypoplasia or both.
- ▶ Has the potential for malignancy transformation and infection.
- ▶ *Prognostic tool: CCAM Volume Ratio(CVR) as three dimensional measurement of lung lesion volume divided by head circumference*

CPAM

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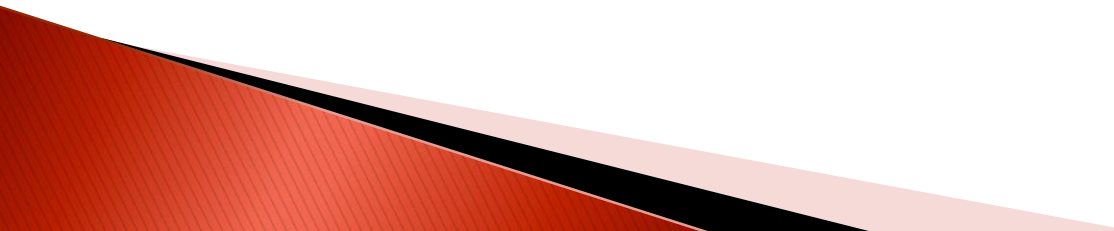
- ▶ *$CVR \geq 1.6$ is in risk of hydrops and in need of fetal interventions*
 - ▶ Microcystic: open fetal surgery
 - ▶ Macrocystic: Thoracoamnion shunt or aspiration
 - ▶ If they are not candidates for fetal surgery: Prenatal steroid in order to decrease size and prevent hydrops
 - ▶ **Type 0:** Rarest, usually lethal, numerous small cysts, originated from trachea and bronchus, associated with cardiac anomaly and pulmonary hypoplasia
- 

- ▶ **Type 1:** Most common, large cysts(1-10 cm)surrounded with small cysts, originated from bronchiols, rarely associated with other anomalies, leads to hydrops and pulmonary hypoplasia due to large size and pressure effect but has a favorable prognosis
 - ▶ **Type 2:** Second most common, originated from bronchioles, small cysts(0.5-2 cm),50% associated with renal agenesis/dysplasia, CHD,BPS
- 

CPAM

7

- ▶ **Type 3:** Originated from alveoli and bronchioles, exclusively in males, **real CCAMs**, small cysts (below 2 mm), does not regress, has pressure effect on mediastinum, heart and cava and leads to polyhydramnious and hydrops
- ▶ **Type 4:** Large cyst(10 cm), asymptomatic even until childhood, originated from distal tree, great prognosis

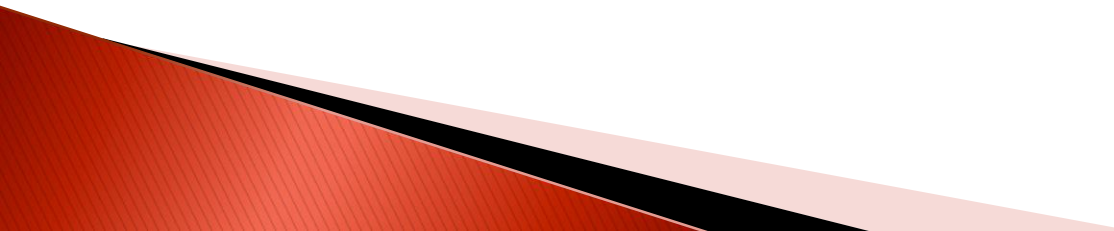
- ▶ CPAM sub-typing in **our study** was as follows:
 - ▶ Type 1 (**5** cases), Type 2 (**2** cases, one of the two was the only hybrid lesion), Type **3** (1 case) and unfortunately **5** cases without specified typing.
 - ▶ Classifying CPAM lesions into macrocystic and microcystic was unfortunately not mentioned in our pathologies. Male gender was significantly more prevalent in these lesions(5 times).
 - ▶ In the study of **Adzick** et al, most of CPAM cases belonged to type 1 category (60-70 %),whereas least common cases were included in type 3 group.
- 

- ▶ Associated anomalies in our study were seen in **three** cases of CPAMs and were as follows:
- ▶ **Mild PS** (in a case of type 1),**Large PDA**(in a case of type 2),**Hypospadiasis** (in a case of undefined type).
- ▶ Of 12 cases of CPAMs, only 4 had prenatal diagnosis.
- ▶ Polyhydramnios was present in only **one** case of type 1.
- ▶ Lesions almost equally involved right and left.
- ▶ Bilateral involvement was detected in only **one** case.
- ▶ There was just **one** female and **one** late preterm case.
- ▶ In the study of **Stocker** et al, **10** of 38 CPAM cases had accompanying congenital anomalies (type 2)

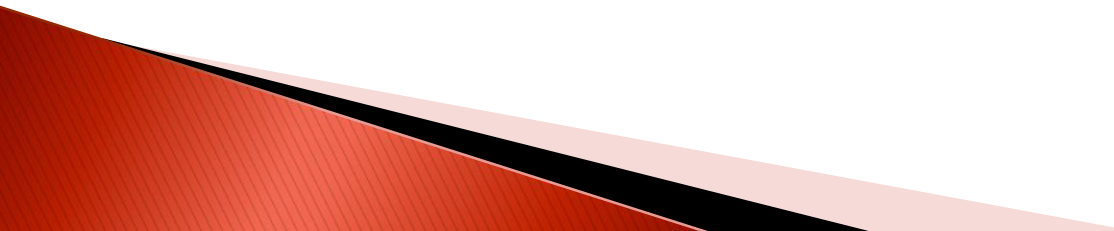
PS

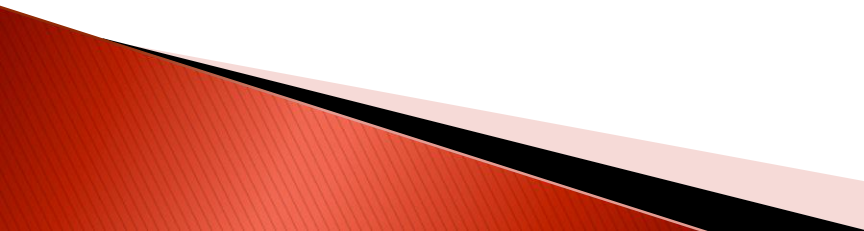
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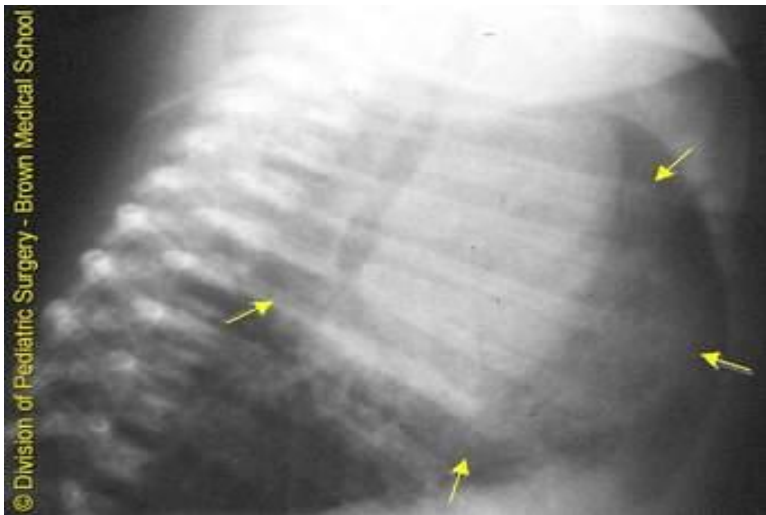
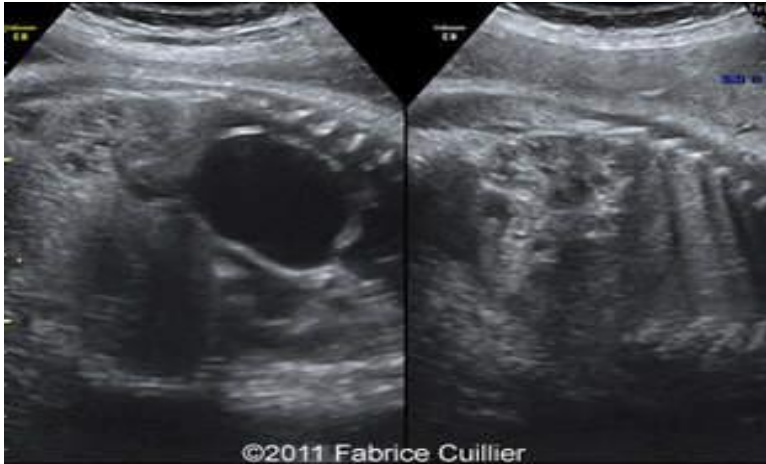


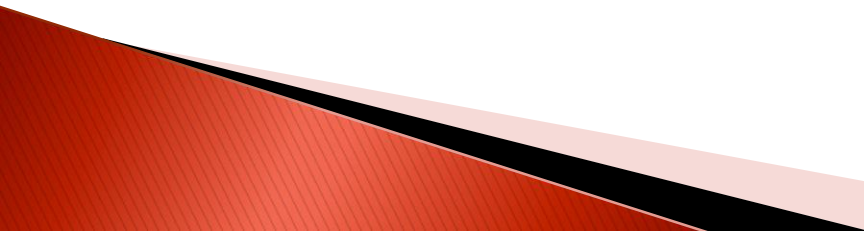
- ▶ Make up to **14-30** % of congenital lung lesions
 - ▶ Classified as extra lobar Sequestration(**ELS**) and intra lobar Sequestration(**ILS**),
 - ▶ More prevalent in male, often located in left lower lobe
 - ▶ ILS is drained into pulmonary venous system.
 - ▶ ELS often has associated malformations(50%) and is drained into systemic venous system.
 - ▶ Vascular supply determination is important for surgical planning.
 - ▶ Heart failure is not unusual due to large left to left shunt.
- 

- ▶ There may be found a history of polyhydramnios.
- ▶ Some of these seem to decrease in size during pregnancy(68%) or disappear post-natally.
- ▶ Have a more favorable outcome than CPAM.
- ▶ *Combination of an aberrant systemic blood supply and an echogenic lung mass in a prenatal ultrasonography is pathognomically in favor of sequestration.*
- ▶ Having considered the common developmentally origin of PS and CPAM, hybrid lesions are not astonishing to appear in pathologies.

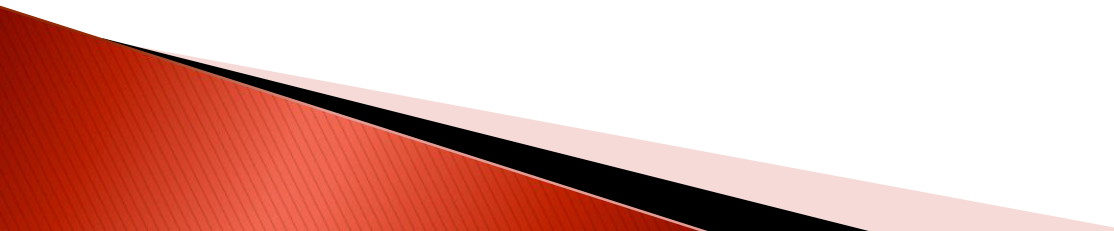
- ▶ In this survey, **3** of 7 cases were diagnosed as ELS,
 - ▶ **Two** BPSs were not defined regarding their subcategory,
 - ▶ **One** case was a hybrid of ELS and CPAM (type 2) which was accompanied by small ASD and PDA. It was detected prenatally as a semi solid mass with cystic lesions.
 - ▶ Only **one** case was an ILS.
 - ▶ Only **two** had prenatal diagnosis
- 

- ▶ The Hybrid lesion was the **only** case for which the blood supply was defined pre-surgically
 - ▶ There were no sex or location preferences.
 - ▶ No accompaniment with polyhydramnios, hydrops and congenital diaphragmatic hernia was detected.
 - ▶ In the study of **Longston** in Texas ELSs were almost **two times** more than ILSs; whereas in the study conducted in **Saudi Arabia** such classification was not mentioned.
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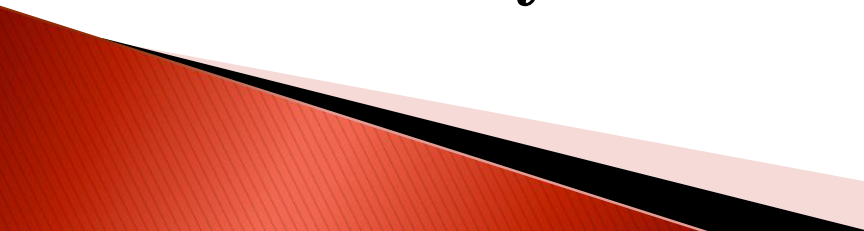


- ▶ Found in mediastinum in up to **2/3rd** of cases
 - ▶ The remainder is found within the lung parenchyma, pleura and diaphragm
 - ▶ Most cases present post-natally, usually with pulmonary infection or hemorrhage
 - ▶ Malignant transformation has not been reported in relation to these lesions.
 - ▶ Our **only** case of right BC was a 6 day old term male with associated findings of prenatal polyhydramnios, congenital diaphragmatic Hernia and jejunal atresia.
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Therapy strategies

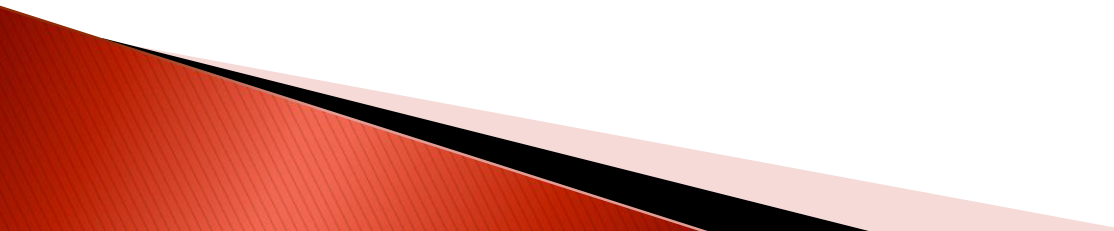
- ▶ Surgeons more or less decide to postnatally resect the lesions through lobectomy or segmentectomy, Although other fetal interventions do exist, for example; amniotic fluid centesis to alleviate polyhydramnios, open fetal surgical resection, thoracoamniotic shunt placement and percutaneous laser ablation .
 - ▶ All of the cystic lesions in our center were post-natally managed with lobectomy.
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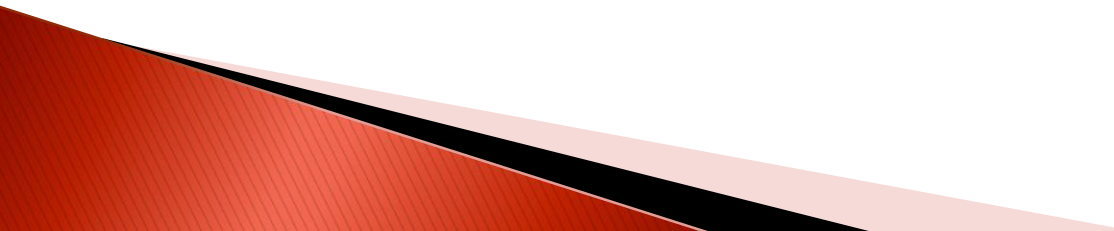
Prognosis


- ▶ Historically polyhydramnios and mediastinal shift were indicators of poor prognosis.
 - ▶ Nowadays, **bilateral lesions, existence of hydrops and smaller cysts** have been proposed as poor prognostic factors.
 - ▶ Overall, prognosis is favorable in respect to surgical lobectomy.
 - ▶ In our survey all of the cases survived and were discharged (length of hospitalization was **between 4 and 37 days** with mean duration of **13 days**).
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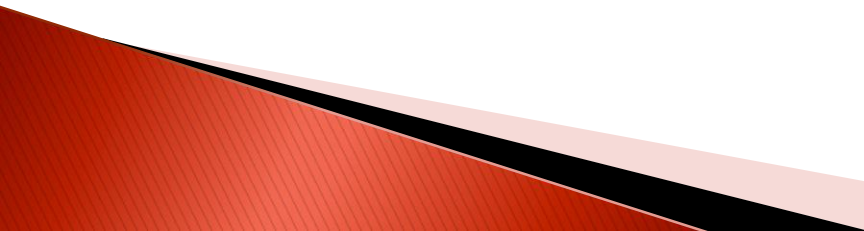
Conclusion

- ▶ **Male** predominance was obvious in the cases just as previous studies.
 - ▶ Unfortunately, **73%** of the cases did not have prenatal diagnosis.
 - ▶ **Actually this statement may be an outstanding warning of either shortage of prenatal care among iranian mothers or lack of sufficient experience or precision in prenatal detection of congenital pulmonary lesions.**
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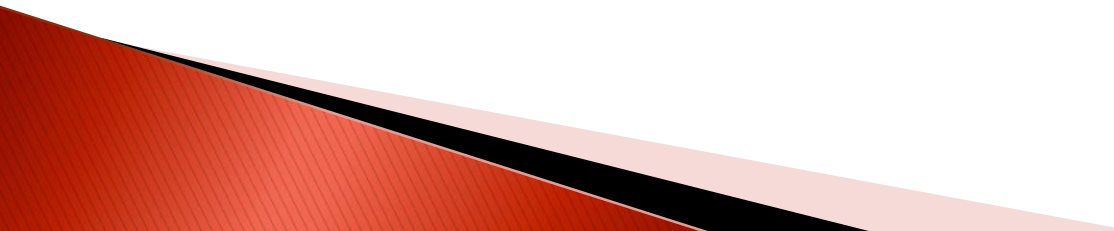
- ▶ Such neglect was more prominent for diagnosis of CLE[55%], in other words most of the cases with undefined prenatal diagnosis were post-natally detected as CLE.
 - ▶ *Lung tissue may be wrongly misdiagnosed as liver or bowel in prenatal sonographies; therefore MRI allows further differentiation when there is suspicion about **CCAM** versus **CDH** (Congenital Diaphragmatic Hernia).*
- 

- ▶ Developed accuracy through **ultra-fast MR** sequences has resulted in exclusion of confounding factors like fetal movement or maternal respiration.
 - ▶ The study could provide us with further results if prenatal screening sonographies were detected and followed post-natally in order to survey the possible course of regression and survival of poor prognostic cases.
 - ▶ Newborns with in time diagnoses would not undergo difficulties of complicated birth, hasty transfer planning and sometimes risky emergent surgeries.
- 

- ▶ Pediatric surgeons should get experience further for **fetal surgical interventions** in order to interfere prenatally for poor prognostic lesions and provide the normal lung development by different techniques of lesion ablations.
 - ▶ **Lack of documented pathology report** of resected pulmonary lesions in 73% of cases was the second most important pitfall which requires reconsideration of the hospital file recording and follow up system.
 - ▶ Based on possibility of malignant transformation of some of these lesions, **immune-histochemical analysis** seem to be necessary to predict such future morbidity.
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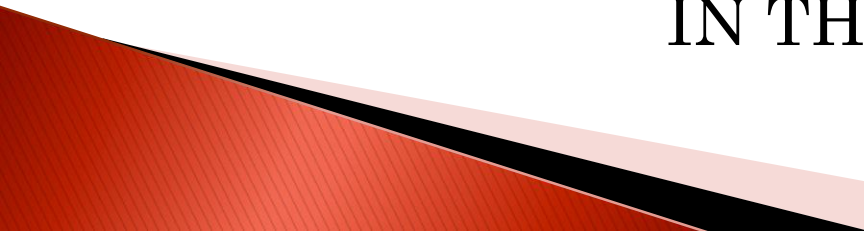
- ▶ The imaging work up of the patients with pulmonary cystic lesions should include a barium swallow when there is a possibility of gastrointestinal communication and also an echocardiography .
 - ▶ **Echocardiography was routinely done during the hospital course but GI Contrast study was performed in none of the cases.**
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In all, we as authors of this study think that the medical experts should always be suspicious of these lesions, follow **a more step-wise approach** and manage the patients as a disciplined team when facing these lesions;



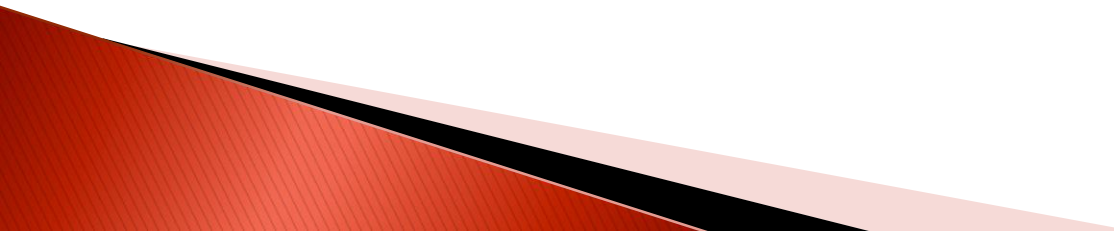
Early and accurate diagnosis of radiologist with the aid of different imaging modalities;
In time prognosis detection of gynecologist and if indicated consultation with a fetal surgeon;
Wise planning for delivery in a tertiary hospital for post natal probable subspecialty care and procedure of neonatologists and continuation of management and evaluation in NICU; and
Precise pathology diagnosis for further follow ups

**ARE THE KEY POINTS
TO BE CONSIDERED
IN THE FUTURE.**



Acknowledgements

We would like to express our gratitude to pediatric surgeons of Mofid Children's Hospital of Tehran for their perfect surgeries and their tireless efforts...





علم را دو پر گمان را یک پرست
ناقص آمد ظن به پرواز ابترست

**Bilginin iki kanadı vardır, şüpheninse tek.
Zan noksandır, uçmaz**

**Knowledge has two wings, Opinion one wing:
Opinion is defective and curtailed in flight**



مرغ یک‌پر زود افتد سرنگون
باز بر پرد دو گامی یا فزون

**Tek kanatlı kuş, çabucak baş aşağı düşer.
Sonra uçmaya savaşıır ama ya iki adımlık
bir yer aşabilir, ya birazcık daha fazla**

**The one-winged bird soon falls headlong;
then again it flies up some two paces or
(a little) more**

افت خيزان می رود مرغ گمان
با یکی پر بر امید آشیان

Şüphe kuşu düşe kalka ümit yuvasına tek
kanatla uçmaya savaşıır

The bird, Opinion, falling and rising, goes on
with one wing in hope of (reaching) the nest

چون ز ظن و ا رست علمش رو نمود
شد دو پر آن مرغ یک پر پر گشود

**Fakat şüpheden kurtuldu da bilgi sahibi
oldu mu o tek kanatlı kuş, iki kanatlı
kesilir. Kanatlarını açar**

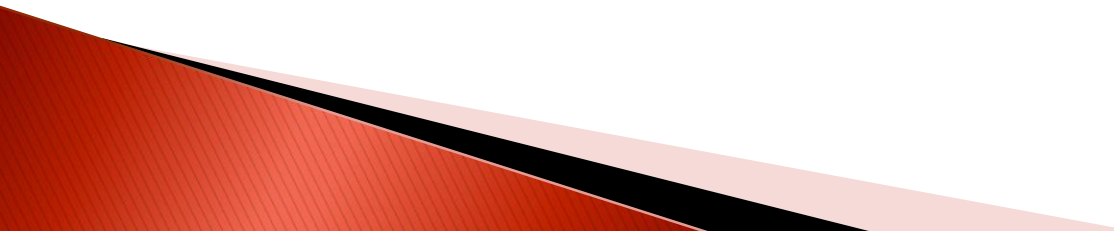
**(But) when he has been delivered from
Opinion, Knowledge shows its face to him:
that one-winged bird becomes two-winged
and spreads his wings**



با دو پر بر می‌پرد چون جبرئیل
بی گمان و بی مگر بی قال و قیل

**Cebraıl gibi iki kanatlı şüphesiz, hilesiz,
kıyl ü kalsiz uçar.**

**He flies aloft with two wings, like Gabriel,
without opinion and without
peradventure and without disputation.**





Thanks for your patience