

Current Concepts in Pediatric Radiology

Edited by O. Eklöf

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With Contributions of

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Preface

The first radiological examination of an infant was reported as early as 1896. This was the prelude to a tremendous amount of pioneer work which was accomplished in the following decades. Interaction of increasing experience, technical improvement and new therapeutic achievement led to the present status of pediatric radiology. In almost any investigation and evaluation of childhood disease, pediatric radiology has a cardinal role.

The establishment of the European Society of Pediatric Radiology in 1963, and the start of the journal *Pediatric Radiology* 10 years later, have created international platforms for the circulation of rapidly accumulating knowledge. Both ventures have helped to underpin high educational standards in the specialty. In many countries national post-graduate courses have contributed quite admirably to favourable trends. More than ever, today medicine is an international science: medical training on an international level has therefore come to stay. This results in a more rapid dissemination of "Current Concepts in Pediatric Radiology" among all those devoted to this fascinating branch of radiology.

This book, mainly based on the lectures given at the First International Post-Graduate Course sponsored by the Swedish Society of Pediatric Radiology in May 1976, presents the contributions of the distinguished guest speakers on selected topics.

It is my belief, as Editor, that this volume will serve as a permanent record and provide interesting reading for those who were unable to attend. For those who had the chance of participating in this meeting the volume will be a useful written statement of the information presented.

Stockholm, September 1977

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Contents

Radiology of Respiratory Distress in the Newborn. A "Gamut of Pattern" Approach (A. GIEDION)	1
1. Introduction	1
2. Preliminary Scan of the "Babygram"	1
2.1 The Main Types of Pattern to be Distinguished	1
3. Conclusion	7
References	11
Chronic Lung Disorders in Childhood (F. N. SILVERMAN)	13
1. Introduction	13
2. Chronic Aspiration Pneumonia	13
3. Congenital Malformations	14
4. Allergic Diseases	16
5. Systemic Diseases	17
6. Chronic Infections	19
7. Conclusion	20
References	21
Therapeutic Embolization of Arteriovenous Pulmonary Fistulas by Catheter Technique (W. PORSTMANN)	23
1. Introduction	23
2. Case Histories	25
3. Discussion	27
4. Summary	30
References	30
Pediatric Gastrointestinal Fiberendoscopy (N. CREMER, S. CADRANEL, P. RODESCH, and M. CREMER)	32
1. Introduction	32
2. Instruments	32
3. Preparation and Premedication	33
4. Training	34

5. Indications and Findings	34
6. Therapeutic Aspects of Endoscopy	40
7. Conclusion	41
References	41

The Esophagus in Infancy (B. R. GIRDANY)	43
References	46

Less Common Disease Patterns in the Gastro-Intestinal Tract with a Special Note on Meconium Ileus (R. ASTLEY)	48
--	----

1. Introduction	48
2. Oesophageal Atresia	48
3. Isolated (H-type) Tracheo-oesophageal Fistula	48
4. Achalasia	48
5. Chronic Granulomatous Disease	48
6. Hiatal Hernia	49
7. Hypertrophic Pyloric Stenosis	51
8. Peptic Ulceration	51
9. Acute Pancreatitis	52
10. Meconium Ileus	54
References	58

Small Intestine — the Terminal Ileum Loop (M. A. LASSRICH)	59
References	65

Current Views on the Diagnosis of Colonic Aganglionosis (B. J. CREMIN, V. BOSTON, D. BROCKWELL)	67
--	----

1. Introduction	67
2. Pathogenesis	67
3. Pathophysiology	67
4. Diagnosis of Hirschsprung's Disease	68
4.1 Clinical Diagnosis	68
4.2 Radiological Diagnosis	68
4.3 Manometric Diagnosis	72
4.4 Histological Diagnosis	72
5. Conclusion	73
5.1 Acknowledgements	74
References	74

Renal Cysts and Cystlike Conditions in Infancy and Childhood (C. FAURÉ)	76
--	----

1. Introduction	76
2. The Multicystic Kidney	76
3. Familial Cystic Renal Conditions	79
4. Miscellaneous Renal Cystic Conditions	84
References	87

Aspects of Acute Kidney Injury in Young Infants (A.R. CHRISPIN) 88

1. Introduction	88
2. The Kidney and the Shock State	88
2.1 Pathological Types of Kidney Injury	88
2.2 Clinical Causes of the Shock State	88
3. Clinical Features and Radiological Findings in Tubular Necrosis and Medullary Necrosis	89
3.1 Acute Illness	89
3.2 Late Studies	90
3.3 Experimental Confirmation of the Urographic Findings	90
4. Clinical Features and Radiological Findings in Renal Venous Thrombosis	91
4.1 Clinical Features	91
4.2 Radiological Findings	91
5. Involvement of other Organs in the Shock State	92
6. Conclusion	92
References	93

The Radiology of the Vesico-Ureteric Junction (H. FENDEL) 94

References	105
----------------------	-----

The Prostate in Pediatric Radiology (G. THEANDER) 106

1. Introduction	106
2. Radiologic Examination	106
3. The Prostatic Tissue	106
3.1 Malformation	106
3.2 Hematoma	106
3.3 Infection	107
3.4 Tumours	107
4. The Prostatic Channels	108
4.1 Anomalous Channels	108
4.1.1. Bowel Fistula	108
4.1.2 Transprostatic Ureter	109
4.2 Normally Existing Channels	110
4.2.1 The Utricle	110
4.2.2 The Ejaculatory Ducts	110
4.2.3 The Glandular Ducts	111
References	114

Approaches to the Evaluation of the Hand in the Congenital Malformation Syndromes (A. K. POZNANSKI) 115

1. Introduction	115
2. Alterations in Configuration of the Hand Bones	115

3.	8	Evaluation of the Size of the Hand Bones	121
3.1		The Metacarpal Sign	121
3.2	8	Bone Length Measurements	122
3.3		Bone Quality	123
3.4	8	Slenderness Ratios	124
4.	8	Determination of Skeletal Maturation	124
5.		Summary	125
		References	125

Lesions of the Spine (G. B. C. HARRIS)

1.		Introduction	127
2.		Vertebral Anomalies — a Useful Clue	127
3.		Scoliosis as a Presenting Sign	129
4.		Back Pain in Children	130
4.1		Infection	130
4.2		Eosinophilic Granuloma	130
4.3		Tumors	130
4.4		Aneurysmal Bone Cyst	131
4.5		Osteoblastoma	131
4.6		Osteoid Osteoma	132
4.7		Trauma	132
5.		Summary	133
		References	134

Hydrocephalus in Children. A Radiological Study (M. HASSAN)

1.		Definition and Classification	135
2.		Distribution of Patients	135
3.		Plain Skull Examination	135
4.		Special Procedures and Results	138
5.		Differential Dignosis	145
6.		Conclusion	147
		References	147

Subject Index

			149
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Radiology of Respiratory Distress in the Newborn

A "Gamut of Pattern" Approach

A. Giedion

1. Introduction

The following "primer" is addressed to colleagues with little experience in this field of neonatal radiology. The identification of typical patterns should be helpful in establishing a list of possible diagnoses (gamut); the final correct choice will usually be guided by the clinical circumstances. Obviously, only an intimate knowledge of physiology, pathophysiology and pathology of the newborn will enable the radiologist to reach a mature judgement. The interested reader is referred to several excellent books and numerous publications for a more extended study of this subject [1, 4, 34, 38, 41].

A first version of our somewhat primitive pattern-gamut approach was published 10 years ago [13] and summarized in the syllabus of the 18th (1975) San Francisco postgraduate course in diagnostic radiology [16]. This method has been, in our hands, particularly helpful for the instruction of our residents working in the newborn unit. Here, fast decisions have to be made at odd times, when an experienced interpreter of the radiologic findings may not be available.

The complex problem of congenital heart disease in the newborn will not be discussed in this paper, and you are referred to some recent publications [19, 22]. Only some implications on the pulmonary pattern, caused by congenital heart disease, will be mentioned. We should, however, keep in mind that a most severe example of congenital heart disease, e.g., transposition of the great vessels with little cross shunt, may have a normal chest, as opposed to a cyanotic baby with pulmonary congestion and an enlarged heart, caused by too high a hematocrit [13].

2. Preliminary Scan of the "Babygram"

Instead of a chest film we prefer, for our initial examination, a "babygram". Changes visible in the skeleton (e.g., syphilis, bone dysplasias), a ruptured spleen, a large liver in congenital heart disease etc., all conditions leading to "respiratory distress", may be recognized at a glance. Also film quality, artifacts, position of the diaphragm etc., have to be checked before concentrating on the chest.

2.1. The Main Types of Pattern to be Distinguished (Fig. 1)

Sometimes, even the decision whether or not a newborn chest is normal or abnormal may be difficult in view of transient normal but quite striking pulmonary patterns [29].

The White (Water Density) Chest (Table 1). Its recognition offers no problem. Obviously, the white chest is the normal radiological appearance before the first breath [11]. Only few of the many causes will be suspected on the evidence of a single film (Table 1). Some additional clinical facts, e.g., bloody foam from the airways or the knowledge of earlier films, showing the reticular granular pattern of hyaline membrane disease, may still allow the correct radiologic diagnosis. Although, quite rare, the immediate recognition of a hydrothorax (Fig. 2) is even more urgent than that of a tension pneumothorax [12].

The Black (Air Density) Chest (Table 2). This group contains only two major diagnoses. Yet, just because these films are apparently typical, they have to be analyzed with particular care.

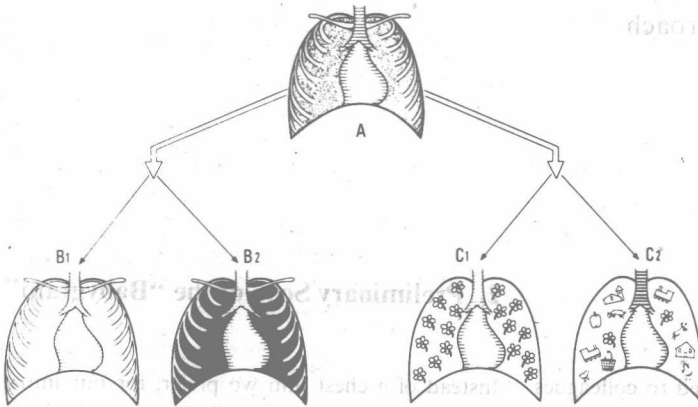


Fig. 1. Schematic drawing displaying the 5 basic "patterns" of the newborn chest. A = normal, B1 = "white"

chest (water density), B2 = "black" chest (air density), C1 = regular pattern, C2 = irregular pattern

Table 1. "White thorax" (water density) of the newborn

Pathogenesis	Cause/Disease
I. Atelectasis	Primary/Secondary > Surfactant (HMD)
II. Alveoli filled or overdistended with fluid	Hypoplastic lung Primary apnea Retention of alveolar fluid (Obstruction) Pulmonary edema Massive aspiration Alveolar hemorrhage Pneumonia
III. Displacement of pulmonary tissue (space-occupying mass)	Tumor (diaphragmatic hernia)
IV. Pleural fluid	Hydro-chylo-hemato-infuso-urinothorax [11a, 12, 23, 35]

Table 2. Black thorax (air density) of the newborn

(Pseudo) pneumothorax
Alveolar emphysema (1st/2nd)
"Cysts"
(Oligemia)

Furthermore, a highly significant *tension pneumothorax* may be missed in the supine newborn, if only "free air" or displacement of the mediastinal structures is looked for. The "free air", sitting as a bubble on top of the lung, may just compress it and change its pattern (flattened—out lung). The "sharp edge sign" [32] is caused by the interface air/mediastinum as opposed to the normal interface lung/mediastinum, causing a more than usually sharp outline of heart and thymus. Both signs allow a preliminary diagnosis. The free sub-sternal air may be demonstrated in a lateral chest film, horizontal beam, supine position of the baby [26]. In our hands, a-p views, the baby lying on its healthy side and with a horizontal beam, have been more diagnostic (Fig. 3).

The most deceiving picture of an *inflated intrathoracic stomach* (Fig. 4) illustrates the need for a careful examination even of seemingly "obvious" cases. Also large cysts, alveolar emphysema and skin-folds may be misinterpreted as pneumothorax (pseudopneumothorax, Table 3). The radiologically quite uniform appearance of *unilateral alveolar emphysema* may be caused by a variety of anatomical factors [21]. A few distinctive features are mentioned in Table 4. In this condition, the

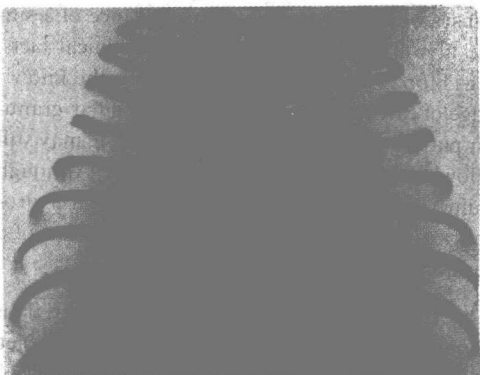


Fig. 2. Hydrothorax. Female 3 h old. Note expanded white chest. Some paraspinal air (artificial respiration). (From [12])

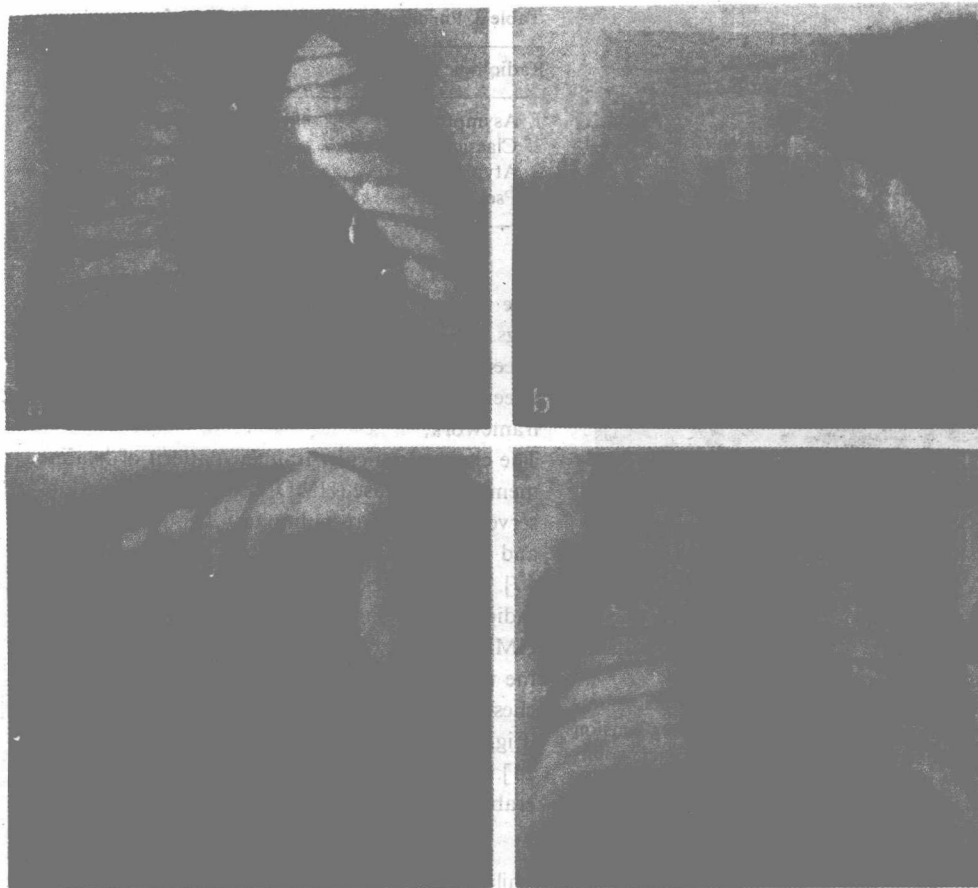


Fig. 3a-d. Female 2 days old. a) On right side typical miliary interstitial emphysema, which is similar, but lighter on left side (pattern change!). b) Lateral view, horizontal beam, with patient in supine position is inconclusive (no free air below sternum visible).

c) A-p view, horizontal beam, patient lying on his healthy side, discloses large amount of air (tension pneumothorax). d) After successful treatment an identical pattern on both sides

involvement of all lobes on one side speaks against the most common idiopathic type, where the lower lobes are usually atelectatic. A few additional typical X-ray findings may offer valuable diagnostic clues: An esophagogram, e.g., may reveal a bronchogenic cyst, which can be missed even at operation with a subsequent unnecessary pneumonectomy [10] (Fig. 5). The radiologic work-up of alveolar emphysema should therefore include an esophagogram and possibly a preoperative bronchogram. In cases where a cardiovascu-

lar factor is suspected (Table 4) angiocardiography may be helpful. Finally, for correct longitudinal interpretation, the three phases of bronchial obstruction have to be understood by the radiologist (Fig. 6) [3, 8, 18].

The Regular Pattern (Table 5). The distinction of a (round) air space and a reticular-interstitial-nodular pattern (Fig. 7) is of course quite arbitrary and mainly didactic.

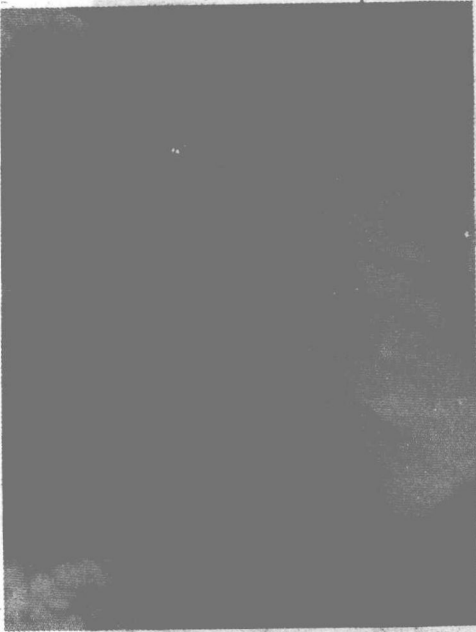


Fig. 4. Female, several hours old, intrathoracic stomach (large diaphragmatic hernia), simulating tension pneumothorax. Note defects in cervicothoracic spine, site of anterior meningomyelocele. (By courtesy of Dr. Bärlocher, Kinderspital St. Gallen)

Table 3. Pneumothorax of the newborn

Radiologic types

1. Asymptomatic
2. Classical tension
3. Atypical tension
4. Pseudo

The Round Air Space Pattern (Table 5A and Figs. 8 and 9). The main distinctive features between the various types is the changing ratio between "air holes" and the surrounding pulmonary framework.

The classical reticulo granular pattern of hyaline membrane disease (HMD) (Fig. 8a) has been observed also in pure pulmonary hemorrhage [25] and β -hemolytic streptococcal disease [3, 38a, 40]. The suggestion of the latter possibility by the radiologist might be life-saving to the patient. HMD disease of course presents radiologically the whole spectrum from near normal to a white chest. *Miliary interstitial pulmonary emphysema* (Fig. 8b,c) has been recognized only recently [6, 24]. Artificial respiration with sometimes considerable peak pressures has contributed to its in-

Table 4. Radiologic differential diagnosis of some types of unilateral alveolar emphysema^a

	Distribution	Radiologic hint	Cause
Idiopathic lobar emphysema	Usually upper and/or middle lobe	Atelectatic lower lobe	Majority idiopathic; rarely intrinsic/extrinsic obstruction
Pulmonary sling [7]	Usually entire right lung, occasionally atelectatic right middle/lower lung	(Total) Right-sided emphysema low left hilus; anterior bowing right main bronchus	Left pulmonary artery arises from right pulmonary artery "sling" around right pulmonary artery
Massive dilatation of pulmonary artery [5a]	Usually right middle or left upper lobe	Emphysema + large hilar density	Congenital avalvular pulmonar artery [5a]; also poststenotic dilatation
Bronchogenic cyst [10] ^b	usually entire right or left side	Unilateral emphysema; broad indentation of esophagus and trachea/bronchus; sometimes mediastinal mass	Bronchogenic cyst

^a Depending on the phase, the emphysema may present radiologically in various ways (Fig. 6).

^b Some cases present in infancy, as in the older child or adult, without respiratory impairment [10].



Fig. 5a and b. Female premature baby, 2 months old.
a) Unilateral hyperlucent lung (alveolar emphysema).
b) Combined tracheobronchography and esophagog-

raphy discloses bronchogenic cyst compressing mainly left bronchus

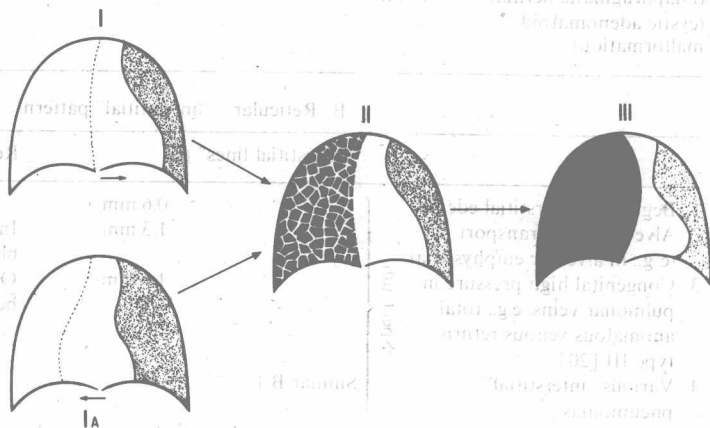


Fig. 6. Alveolar unilateral emphysema in newborn. Schematic drawing of various phases. *I*. Retention of intraalveolar fluid, space-occupying white mass. *IIA*. Same as *I*, but combined with some diminished vol-

ume of the affected lung. *II*. Transportation phase of alveolar fluid through lymphatics (reticular pattern; see Fig. 10c). *III*. Classic unilateral hyperlucent, over-expanded lung

creasing frequency. This finding must alert the physician to further complications (tension pneumothorax, pneumopericardium, etc. (Table 6). Again, a spectrum of four radiologic main patterns can be observed [24] (Table 5, Fig. 3, 8b-d, h). *Isolated pulmonary lymphangiectasis* may present as a round airspace pattern (Fig. 8g). In at least one case [14] associated hypoplasia of the bronchial cartilage might have been a contributory factor to the airway dilatation. This pattern,

also described as reticulonodular [36], has been seen only in primary, isolated, congenital lymphangiectasis (group III of Noonan [30]): Apparently however, some cases just show the well-known pattern of interstitial pulmonary edema [36].

Cystic adenomatoid malformation may show a very similar, yet unilateral pattern: Again, this histologic diagnosis encompasses the whole spectrum from these multicystic lesions (73.3%) (Fig. 9a), to

Table 5*

A. Round air space pattern

	a) Airspace Ø ^a	b) Relation wall Ø ^a to air space Ø.	Remarks
Hyaline membrane p. classic reticulo granular p.	0.6–1.0 mm	> a	Dynamic changes to other stages [15]; same pattern also in pulmonary hemorrhage and early onset streptococcal infection
Interstitial miliary emphysema	circa 1 mm	≥ a	History of positive pressure breathing; other patterns: pseudo-cystic-bullous-linear
Pulmonary (isolated) lymphangiectasis	circa 2 mm	< a, Also patches	Survival only a few days or weeks
Cystic adenomatoid malformation		< a, Also patches	Unilateral
Mikity-Wilson syndrome stage I and broncho-pulmonal dysplasia stage III (Diaphragmatic hernia) (cystic adenomatoid malformation)	1.5 mm + 10 mm +	< a < < < a	Perinatal history (premature/respirator) Sunken-in abdomen, etc.

B. Reticular—"interstitial" pattern

	"Interstitial lines" Ø ^a	Remarks
1. Beginning interstitial edema	0.6 mm	
2. Alveolar fluid transport (e.g. in alveolar emphysema)	1.3 mm	Initially white lung, later black lung
3. Congenital high pressure in pulmonary veins, e.g., total anomalous venous return type III [20]	1.6 mm	Often normal heart size. May become patchy, later alveolar edema
4. Various "interstitial" pneumonias	Similar B I	

C. Irregular Patterns

Coarse irregular pattern (fetal aspiration syndrome)	Coarse patchy irregular infiltrates, 0.5 × 0.5 mm → 9 × 3 mm → confluent local and/or general overinflation	Typical spectrum. Rapid clearing in few days. Typical perinatal history.
Various neonatal pneumonia patterns		Various pneumonias and pulmonary hemorrhage may display similar features

Modified from [16].

These are approximate, average figures from a few cases. They should not be considered as Scientific, accurate data, but of qualitative character.

dominant cysts in multicystic background (13.3%) and to solid homogeneous masses (13.3%) [27].

The Mikity-Wilson pattern stage I [28] (Fig. 8e) and broncho-pulmonary dysplasia stage III ("cystic appearance" [31] (Fig. 8f) may look identical:

We have observed the latter developing as early as the 3rd day of life, obviously still in the neonatal phase.

The Reticular Interstitial-Nodular Pattern (Table 5B, Figs. 10 and 11). The pure interstitial pat-

Table 6. Clinical significance of dx interstitial pulmonary emphysema

Direct:	Stiffening of lung/space-occupying lesion
Warning sign of:	Tension-pneumothorax?
	Pneumopericardium?
	Air embolism? [37]
	Potter syndrome?

tern caused by noninfectious edema and pulmonary infection may look radiologically alike (compare Figs. 10 and 11). The "fluid transportation phase" (see above) of alveolar emphysema is particularly impressive (Figs. 6, 10, c). Interstitial edema in cases of increased pulmonary venous pressure [20] may be misunderstood as infection. This holds particularly true for total anomalous pulmonary venous return below the diaphragm, type III [9], as the heart in these cases may be of normal size and cyanosis may still be absent [20] (Fig. 10b).

The various types of regular patterns indicative of pneumonias again show the full spectrum from an early interstitial edema-like network (Fig. 11a) to a coarse granular (Fig. 11b) and an irregular patchy pattern (Fig. 11e). Bomsel et al. [5] recognize a "quite typical symmetrical pattern of blurred, alveolar opacities" (nodular), the "inversion of the miliary interstitial emphysema pattern," indicative of hematogenous dissemination of streptococcal infection. In our experience, this pattern is also seen in other types of infection, e.g. listeria (Fig. 11b), and the clinician should be alerted to secure material from stomach, rectum, lumbar fluid and blood, to come to the correct bacteriologic diagnosis.

The Irregular Pattern (Table 5c, Fig. 11c-e). The classical "coarse, irregular pattern" [33] (Fig. 11d) of the fetal meconium aspiration syndrome is part

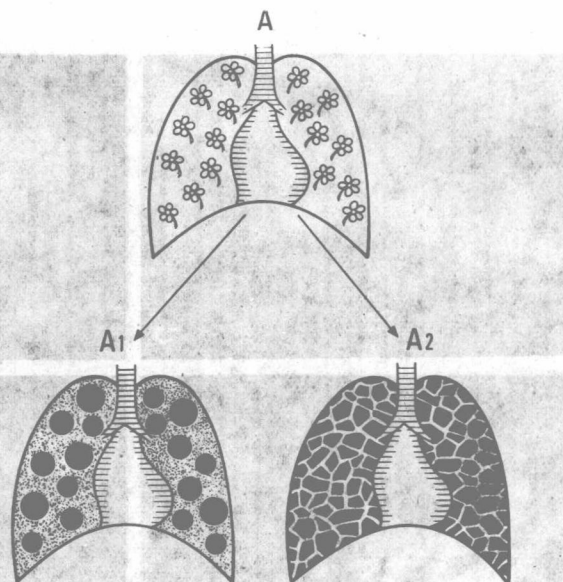


Fig. 7. Schematic drawing of "regular patterns" (A) which may be divided in round-airspace patterns (A1) and reticular-interstitial-nodular patterns (A2)

of a wide spectrum, ranging from minimal findings to the white chest [17].

Pneumonia, hemorrhage, and atelectasis may all contribute to a highly irregular pattern in a chest film. The correct diagnosis will depend largely on additional clinical information, as well as the radiological sequence of events with its change of patterns.

3. Conclusion

Our diagnostic approach is necessarily a static one. Quite often, the four-dimensional approach will offer valuable diagnostic hints. Finally, all enumerated and additional factors do not act in a vacuum. Sometimes, our diagnosis will, at best, be but an educated guess. Still, let's try to be radiologists first!