Current Concepts in Pediatric Radiology

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Dr. Alan R. Chrispin
The Hospital for Sick Children,
X-Ray Department
Great Ormond Street
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England

Volume Editor:
Dr. Ole Eklöf
Röntgenavdelingen, Barnkliniken
Karolinska Sjukhuset
10401 Stockholm 60
Sweden

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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 sponsoned 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

OLE EKLÖF

List of Authors

Dr. R. Astley
Deparment of Radiology
The Children's Hospital
Birmingham, B16 8ET, England

Dr. S. Cadranel
Gastro-enterological Unit
Departement Radiologie Pediatrique
Hôpital Saint-Pierre
Rue Haute, 322
B-1000 Bruxelles

Dr. A.R. Chrispin
The Hospital for Sick Children
X-Ray Department
Great Ormond Street
London, W.C. 1N 3JH, England

Dr. M. Cremer
Department of Gastro-enterology
Hôpital Universitaire Brugmann
B-1000 Bruxelles

Dr. N. Cremer
Departement Radiologie Pediatrique
Hôpital Saint-Pierre
Rue Haute, 322
B-1000 Bruxelles

Prof. B. J. Cremin
Dr. V. Boston
Dr. D. Brockwell
University of Cape Town
Department of Radiology
Groote Schuur Hospital
Observatory, 7925, South Africa

Dr. C. Fauré
Service de Radiologie
Hôpital Trousseau
8-28 avenue de Docteur-Arnold-Netter
F-75571 Paris Cedex 12

Dr. Fendel
Kinderklinik der Universität
München im Dr. von Haunerschen
Kinderspital
Lindwurmstraße 4
D-8000 München 2

Prof. Dr. A. Giedion
Abt. Radiologie
Kinderspital Zürich
Steinwiesstraße 75
CH-8032 Zürich

Dr. B. R. Girdany
University of Pittsburgh
School of Medicine
Department of Radiology
Pittsburgh, PA 15261, USA

Dr. G. B. C. Harris
The Children's Hospital Medical Center
300 Longwood Avenue
Boston, Massachusetts 02115, USA

Dr. M. Hassan
Service de Radiologie
Hôpital Saint-Vincent-de-Paul
74, avenue Denfert-Rochereau
F-75014 Paris

Prof. Dr. M. A. Lassrich
Universitäts-Kinderklinik
Röntgenabteilung
Martinistr. 52
D-2000 Hamburg 20

Prof. Dr. Sc. med. W. Porstmann
Abt. für radio-vaskuläre Diagnostik
Humboldt-Univ. zu Berlin
Schumannstr. 20—21
DDR-104 Berlin

Dr. A. K. Poznanski
Division of Pediatric Radiology
C.S. Mott Children's Hospital
Univ. of Michigan Medical Center
Ann Arbor, Michigan 48101, USA

Dr. P. Rodesch
Gastro-enterological Unit
Department Radiologie Pediatrique
Hôpital Saint-Pierre
Rue Haute, 322
B-1000 Bruxelles

Dr. F. N. Silverman
Department of Radiology
Stanford University Medical Center
Stanford, California 94305, USA

Dr. G. Theander
Department of Radiology
Malmö Allmänna Sjukhus
S-214 01 Malmö

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Radiology of Respiratory Distress in the Newborn

A "Gamut of Pattern" Approach

A. Giedion

1. Introduction

The following "primer" is adressed 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.

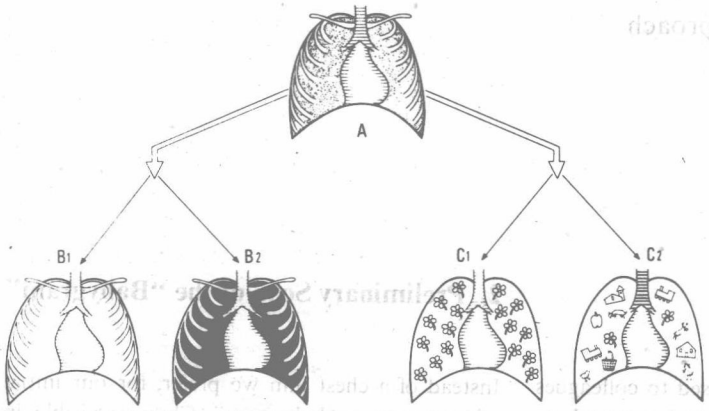

Fig. 1. Schematic drawing displaying the 5 basic "patterns" of the newborn chest. A = normal, B1 = "white" chest (water density), B2 = "black" chest (air density), C1 = regular pattern, C2 = irregular pattern

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

Pathogenesis	Cause/Disease
I. Atelectasis	Primary/Secondary > Surfactant (HMD) Hypoplastic lung
II. Alveoli filled or overdistended with fluid	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]

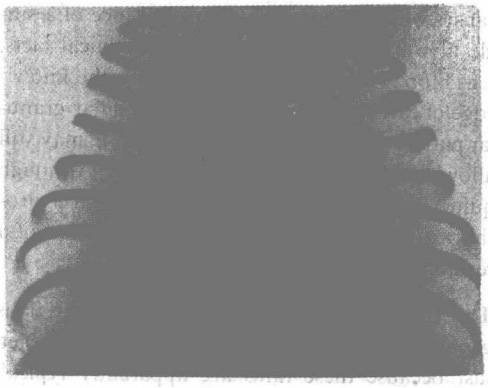

Fig. 2. Hydrothorax. Female 3 h old. Note expanded white chest. Some paraspinal air (artificial respiration). (From [12])

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 substernal 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

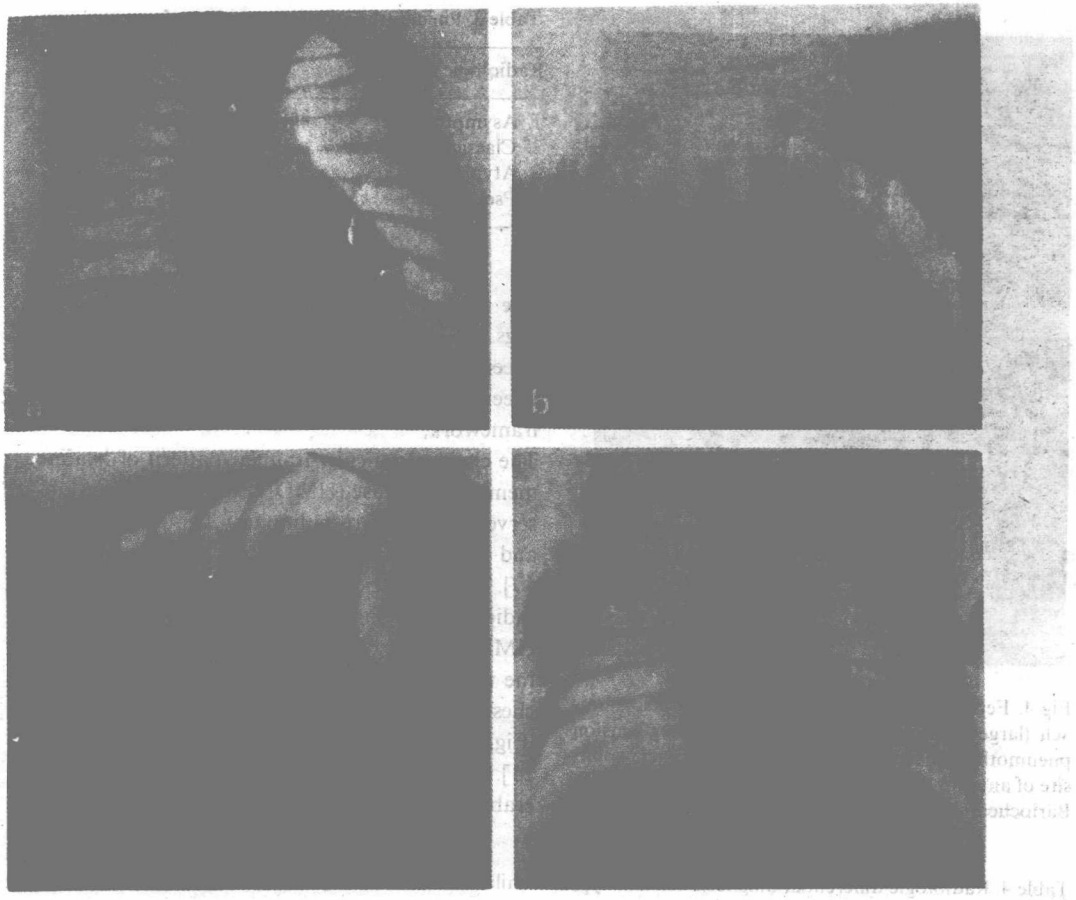

Fig. 3a–d. Female 2 days old. a) On right side typical miliary interstitial emphysema, which is similar, but lighter on left side (pattern change!). b) Lateral view, horizontal beam, with patient in supine position is inconclusive (no free air below sternum visible). c) A-p view, horizontal beam, patient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

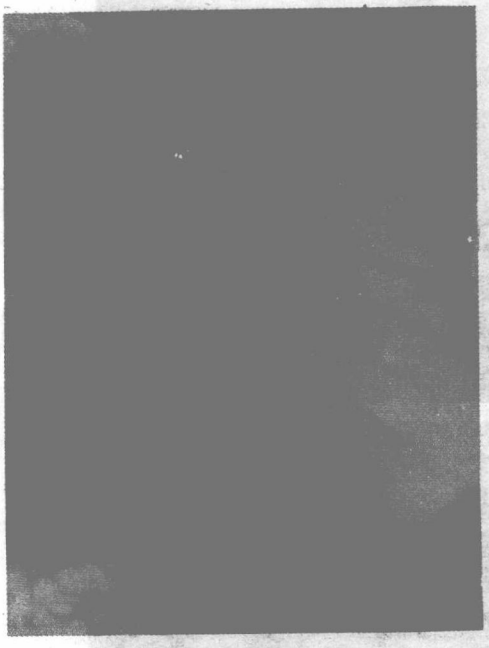

Fig. 4. Female, several hours old, intrathoracic stomach (large diaphragmatic hernia), simulating tension pneumothorax. Note defects in cervicothoracic spine, site of anterior meningomyelocele. (By courtesy of Dr. Bärlocher, Kinderspital St. Gallen)

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].

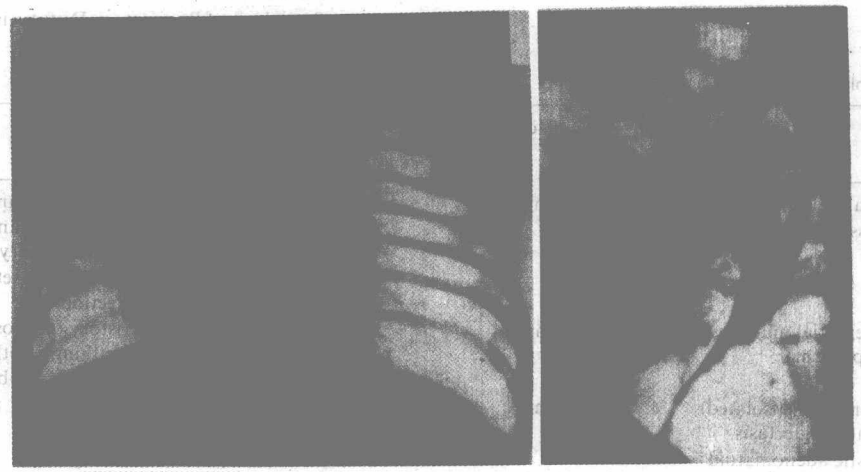

Fig. 5a and b. Female premature baby, 2 months old. a) Unilateral hyperlucent lung (alveolar emphysema). b) Combined tracheobronchography and esophagography discloses bronchogenic cyst compressing mainly left bronchus

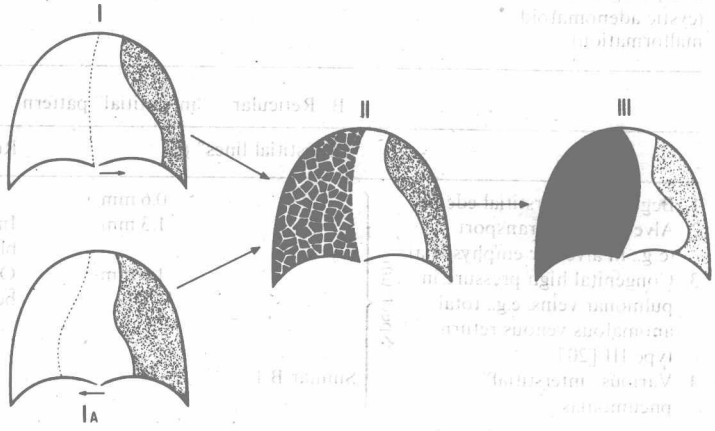

Fig. 6. Alveolar unilateral emphysema in newborn. Schematic drawing of various phases. *I*. Retention of intraalveolar fluid, space-occupying white mass. *IA*. Same as *I*, but combined with some diminished volume of the affected lung. *II*. Transportation phase of alveolar fluid through lymphatics (reticular pattern; see Fig. 10c). *III*. Classic unilateral hyperlucent, overexpanded 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 ∅[a]	b) Relation wall ∅[a] to air space ∅	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	1.5 mm +	< a	Perinatal history (premature/respirator)
(Diaphragmatic hernia) (cystic adenomatoid malformation)	10 mm +	< < < < a	Sunken-in abdomen, etc.

B. Reticular—"interstitial" pattern

	"Interstitial lines" ∅[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 pulmonar 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 1	

Spectrum

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].

[a] 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 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.

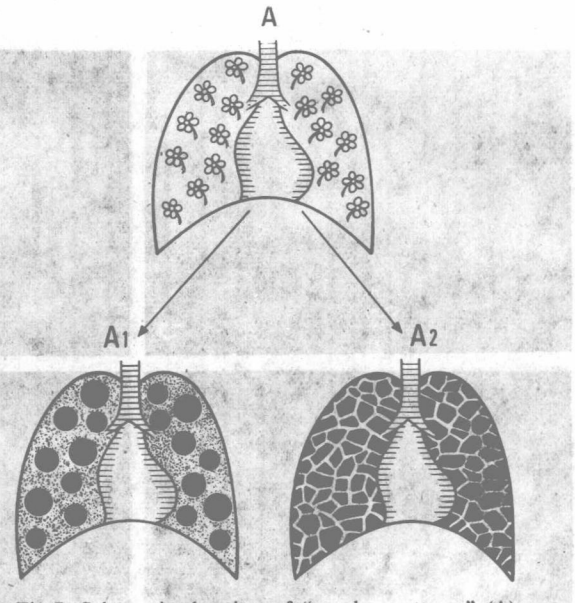

Fig. 7. Schematic drawing of "regular patterns" (A) which may be divided in round-airspace patterns (A 1) and reticular-interstitial-nodular patterns (A 2)

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!