Pediatr Radiol (1982) 12: 57–61

Pediatric Radiology

Originals

Chest Radiographs in Cystic Fibrosis. A Follow-up Study with Application of a Quantitative Scoring System

J. M. van der Put, M. Meradji, D. Danoesastro and K. F. Kerrebijn

Departments of Pediatric Radiology and Pediatrics, Subdepartment of Respiratory Diseases, Erasmus University Medical School, Sophia Children's Hospital, Rotterdam, The Netherlands

Abstract. A modified Chrispin-Norman score was used to investigate at what age typical X-ray signs of cystic fibrosis (CF) appear and for the follow-up of patients with CF. Scores of patients with chronic obstructive lung disease (COLD) and with innocent heart murmurs were used for comparison. The findings were: 1. Hyperinflation and line shadows are early but not specific symptoms of CF. 2. Mottled and ring shadows can be considered as specific for CF. In most patients they only appear after the age of 3. They are never present in COLD. 3. The mean annual increase between the ages of 3–8 is 2 points. No difference existed between boys and girls in total scores and annual increase up to the age of puberty. 4. CF patients with portal hypertension have the same scores as CF patients without portal hypertension.

Key words: Cystic fibrosis – X-ray – Progression – Follow-up

The prognosis of cystic fibrosis (CF) has improved in the last decades and survival rates are increasing with modern therapy [1]. As a result we are now able to follow patients up over longer periods. During this follow-up methods are needed by which the condition of the patient can be assessed.

Shwachman [2] in 1958 was the first who determined the severity of the disease in individual patients in a quantitative manner, in order to compare the results of different therapeutic regimens [3–5]. He applied a numerical score for both clinical data (general activity, physical examination and nutrition) and X-ray findings.

Cooperman [6] proposed a scoring system analogous to the Apgar score for the newborn, in which attention was given to complications like portal hypertension and cor pulmonale.

In 1974 Chrispin and Norman [7] described a scoring system for chest radiographs (C-N score). Attention was given to five roentgen signs, reflecting various pathological processes in the lung. Matthews et al. [8] reported a good correlation between the X-ray scores obtained in this way and pulmonary function tests. The C-N scoring system is used in a cooperative prospective study in Europe on the effect of continuous versus intermittent antibiotic treatment.

The purpose of this study was twofold:

1. To investigate at what age typical signs of CF appear on the chest radiographs in young children.
2. To compare the C-N score at various ages in patients with CF, chronic (recurrent) obstructive lung disease (COLD) (i.e. asthma, asthmatic bronchitis) and children without respiratory disease in whom a chest X-ray was done because of a heart murmur.

We found that in most children after the age of 3 years X-ray findings in CF could be differentiated from those in COLD. After that age the scoring system seemed valuable during the follow-up of patients with CF.

Material and Methods

311 chest radiographs of 90 patients with a proven CF (48 males and 42 females) were studied. The X-rays taken were at approximately yearly intervals. From the first 2 years of life however, 2 or even 3 X-rays were chosen. All patients were treated in the Dept. of Respiratory Diseases of the Sophia Children's Hospital. The ages of the patients on admission ranged from 0 to 17 years, with a peak in the first year. The follow-up period showed much variation. In 42 patients it was either 1 or 2 years. In 20, the follow-up period was 3 or 4 years. In 24 the follow-up was 5, 6 or 7 years and in 4 patients X-rays were available over a longer period.

In the assessment of the chest X-rays we used the scoring system as described by Chrispin and Norman [7], with a small modification that adds four points at the most to the original score. The total score *(s)* is the algebraic sum of the partial scores, assigned for the configuration that is typical for hyperinflation *(a)*, bronchial line shadows *(b)*, mottled shadows *(c)*, ring shadows *(d)*, large shadows *(e)* and – in addition to the original C-N score – for enlarged hilar shadows (e^l), according to the following simple formula: $s = a + b + c + d + e + e^l$. In this formula the score a (hyperinflation) is again composed of 3 partial scores, i.e. sternal bowing (a_1), dia-

0301-0449/82/0012/0057/$01.00

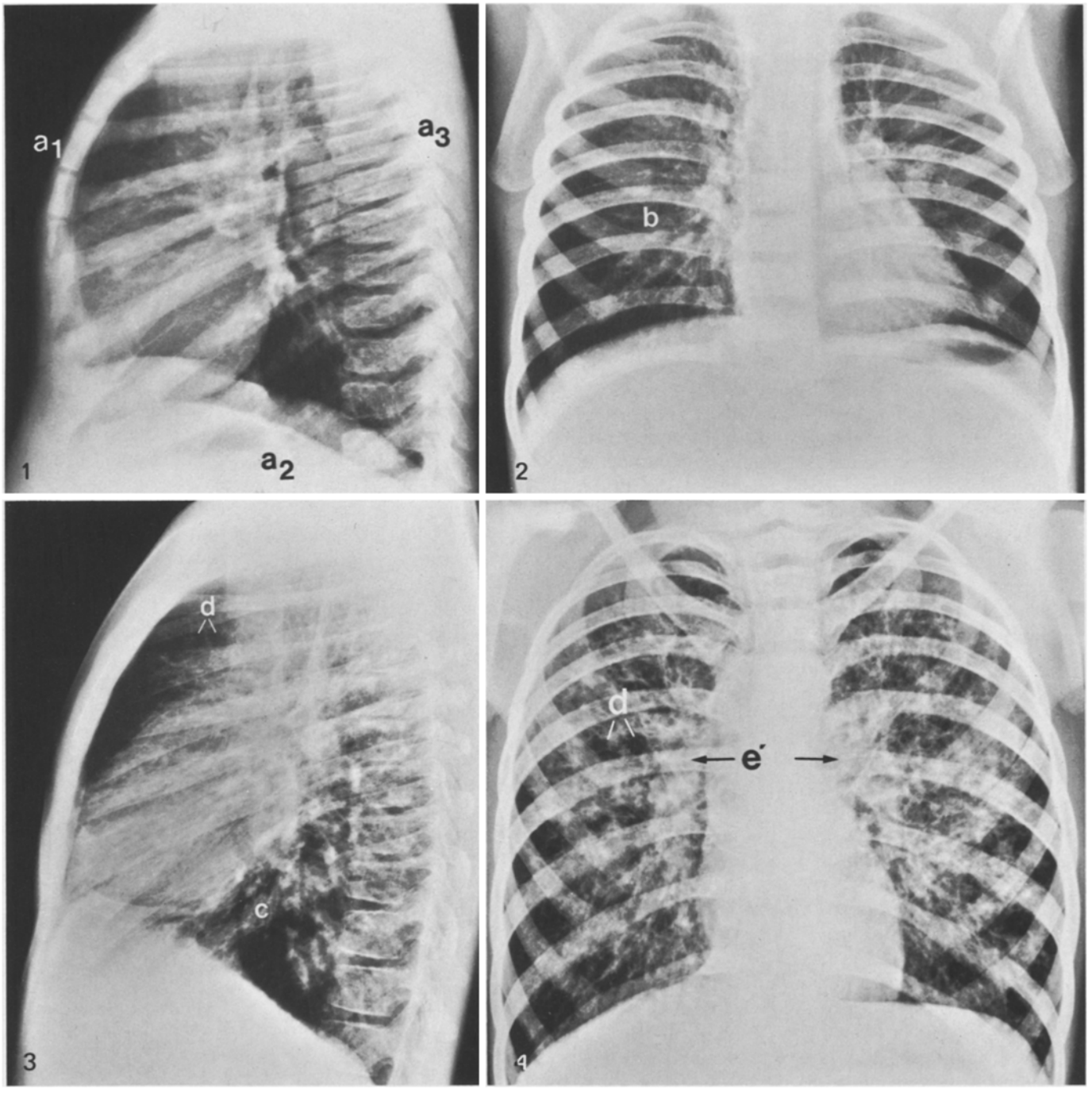


Fig. 1–1. Patient A, lateral chest X-ray. A 4-year-old child with cystic fibrosis. Hyperinflation is illustrated by sternal bowing *(a_1)*, diaphragmatic depression *(a_2)* and thoracic kyphosis *(a_3)*

Fig. 1–2. Patient A, p. a. chest X-ray. Bronchial line shadows *(b)* are seen in a hyperinflated chest

Fig. 1–3. Patient B, a young adult with cystic fibrosis. The lateral view shows signs of hyperinflation and also mottled shadows *(c)* and ring shadows *(d)*

Fig. 1–4. Patient B, p. a. chest X-ray. Enlarged hilar shadows *(e^1)* are present with typical ring shadows *(d)*

phragmatic depression *(a_2)* and thoracic kyphosis *(a_3)*. *($a = a_1 + a_2 + a_3$)* (Fig. 1 a). Twenty patients with COLD, with a history of cough and wheezing at least once a month, in whom at least three X-rays were made with an interval of 1 year or more, were selected for comparison and the same scoring system was applied to their X-rays. It appeared that no patients above the age of 10 were available with three successive X-rays.

In order to obtain chest X-rays of children without respiratory disease we collected the films of a group of 20 individuals aged 1 to 12 years, in whom a chest X-ray was done because of a heart mur-

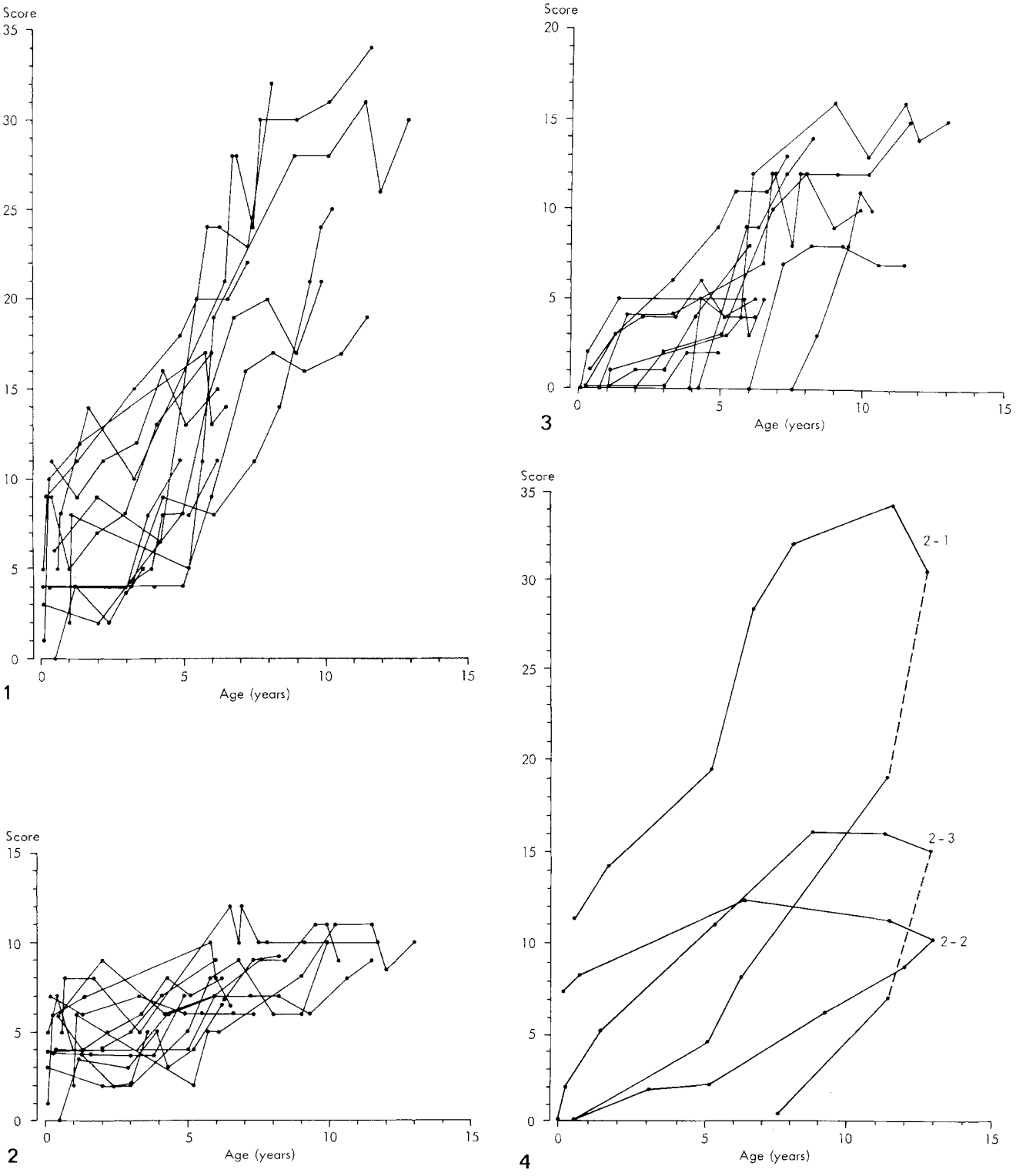


Fig. 2–1. Scores in 13 patients with an early diagnosis (before 1 year of age) of cystic fibrosis, followed up for 5 years or more

Fig. 2–2. Sum of the partial scores *a* and *b* (for hyperinflation and bronchial lines) in the same patient group *(a + b)*

Fig. 2–3. Sum of the partial scores *c* and *d* (for mottled and ring shadows), in the same patient group *(c + d)*

Fig. 2–4. Early diagnosis group. Areas of total scores compared with areas of partial scores *(a + b)* and *(c + d)* known from Figs. 2–1, 2–2 and 2–3

mur that later turned out to be innocent. In this group – as was to be expected – the score was always zero. All X-rays were scored by the radiologists M.M. and D.D.

Results

The earliest signs of cystic fibrosis on the chest X-ray are hyperinflation *(a)* and bronchial lines *(b)* (Figs. 1). These signs are not specific, since they also occur in severe cases of COLD. A striking fact is that mottled shadows *(c)* or ring shadows *(d)* are hardly seen before the age of 3. Once they can be recognized, it is possible to make a certain diagnosis of CF (Figs. 1–3 and 1–4). In infants with CF sternal bowing (a_1) and diaphragmatic depression (a_2) mostly occur together and are as frequent as in the higher age groups. In the first and second year of life only a_1 and a_2 were seen, but later also thoracic kyphosis (a_3) occurs. The scores for a_1 and a_2 showed a temporary small regression (by one or two points) in only four out of 19 children. Thoracic kyphosis (a_3) never showed any regression. This indicates that the individual pattern of hyperventilation signs, once established, remains more or less constant.

Fig. 2–1 shows the total scores of all children known from the first year of life to have CF. For each individual the scores at different ages are plotted and interconnected with lines. There is an obvious tendency to increase with each year, but within a certain range. Between 0 to 3 years of age the scores are rather low (0–15 points). Thereafter the scores gradually increase with 0 to 4 points per year, with a mean of two points per year. If the increase is more or less linear the score will come near to 30 points at the age of 12.

In Fig. 2–2 the partial scores are graphed for hyperinflation *(a)* and bronchial lines *(b)*. This figure is very similar to the graph of the children with COLD (Fig. 3) in whom the scores are also mainly due to hyperinflation of the chest and to bronchial lines.

In Fig. 2–3 the scores for mottled shadows *(c)* and ring shadows *(d)* are shown. The figure illustrates that in most children, but only from about 3 years of age, specific roentgenologic signs of the disease – mottling and ring shadows – can be expected to be present. At this age the median value of the partial scores for these specific lesions has come to a value of two points.

In Fig. 2–4 the areas of Fig. 2–2 and 2–3 are compared. One may consider the total score to be built up from two main components, the one beginning at a higher level but with a slower increase *(a + b)* and the other starting at zero but increasing faster with age *(c + d)*.

In Fig. 3 the scores of the children with COLD are shown. It is important to notice that no mottled or ring shadows are present in this group. This explains that the total scores are lower than in CF and that the partial scores for hyperinflation are identical to the total scores. Some hilar enlargement is found in most patients. The scores have a tendency to increase with age (1 or 2 points per year) but do not become as high as in CF. The maximal score was 13 points.

Fig. 4–1 shows the scores of the children known from the second year of life on or later to have CF. Only this group contains in-

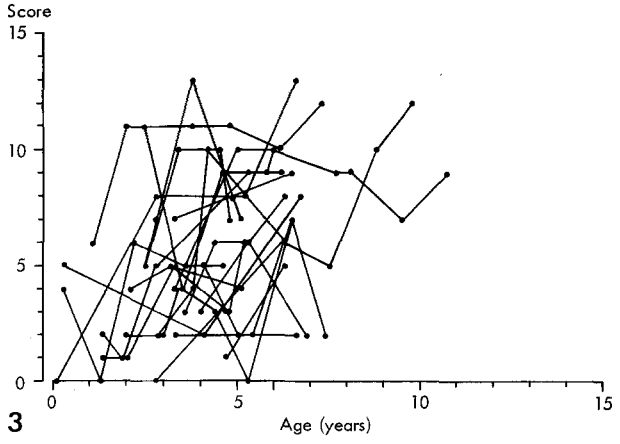
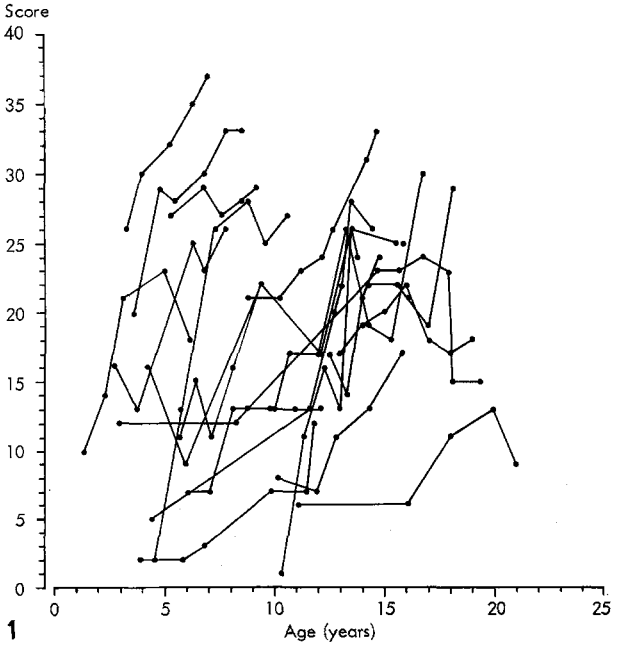


Fig. 3. Total scores of 20 patients with COLD



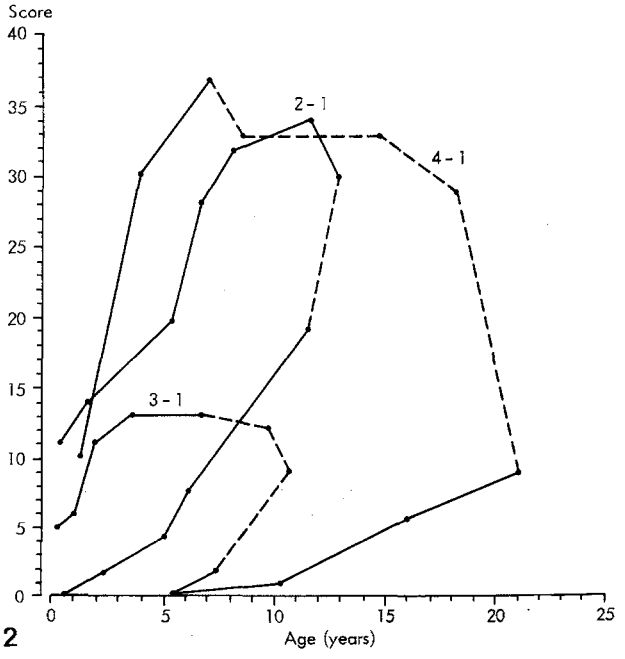


Fig. 4–1. Late diagnosis group. Scores of patients, who were followed up over 5 years or more. There appears to be a strikingly high range in the scores of this group

Fig. 4–2. Areas are delineated of patients from the early diagnosis group (Fig. 2–1), compared to patients with COLD (Fig. 3–1) and patients from the late diagnosis group (Fig. 4–1)

dividuals up to 20 years of age and more. The range of scores is very large and the uniformity in annual increase seems to be less than in the previous group of children with CF. It is very hard to draw conclusions from these data which are derived from a probably very inhomogeneous population.

In Fig. 4–2 the areas are delineated of the three groups of patients, the early diagnosis CF group, the late diagnosis CF group and the COLD group. In the reference group of "normal" chest X-rays the scores were all zero, as mentioned above.

There were no significant differences between boys and girls in age distribution, scores or annual increase in scores (Wilcoxon two sample test). It has been mentioned that in CF a predilection exists for the upper quadrants. We therefore compared the frequency of abnormalities in upper and lower quadrants: 34 out of 80 patients without portal hypertension had more changes in the upper than in the lower quadrants; in 22 the distribution was equal and in 24 more changes were found in the lower quadrants. The differences between these groups are not statistically significant (rank sign test). The ten patients with CF and portal hypertension diagnosed because of an enlarged spleen and esophageal varices, were comparable to the whole population of patients in regard to age and scores (rank sign test). Three had more changes in the upper quadrants, in two the changes in both quadrants were equal and in five the lower quadrants were more affected.

Discussion

Bronchial line shadows and hyperinflation of the lungs are not specific for CF. They can be regarded as early signs of bronchial disease of various causes. The scores in the population of patients in whom the diagnosis CF was made in the first year of life behave differently from the scores in the group of CF patients in whom the diagnosis was made after this age. This may indicate that these two groups contain patients with a difference in severity of CF. However, the overlap between the groups is large.

Looking at X-rays of the same CF patient, differences in the consecutive pictures and variations in the corresponding scores can be noticed. It is very likely that these represent variations in the general condition. It has been noticed that mottled shadows can change into ring shadows when the condition of the patient improves, for instance after a period of intensive treatment. At the same time signs of hyperinflation tend to diminish. As a result the total score will be lower. The reverse happens, if the general condition deteriorates. This results in the individual scores having a range, which reflects changes in general condition. After the age of 2 or 3 years the progression is approximately linear in most patients.

Girls often die from CF at an earlier age than boys. It could therefore be expected that differences would exist in scores and annual increase in scores between boys and girls. However no such differences were found up to the age of puberty. The number of observations at later ages was insufficient to examine this.

Enlarged hilar shadows may result from pulmonary hypertension, in which case they are an ominous sign in a terminal stage. More often, however, enlarged hilar shadows are a sign of lymphadenopathy, occuring in CF with recurrent and long-standing respiratory infections.

It may well be that the use of a score restricted to the presence of severity of mottled and ring shadows without the contribution of hyperinflation and line shadows is as useful as the present score for the follow-up of CF patients.

Conclusions

1. Hyperinflation and line shadows are early but not specific signs of CF. Sternal bowing is generally the first symptom of hyperinflation. Signs of hyperinflation seldom show regression.
2. Ring and mottled shadows are specific for CF and are not present before the age of 3 years in most patients. They are never present in COLD.
3. Between 3 and 8 years of age the mean annual increase is about two points. After the age of 9 the annual increase shows large differences, leading to scores with an increasingly high range.
4. Boys and girls have the same total scores and annual increases up to the age of puberty.
5. Scores from CF patients with and without portal hypertension are similar. In the latter patients we found a slight predilection for predominating lesions in the lower quadrants.

Acknowledgement. We express our thanks to Prof. Dr. R. van Strik for his help with statistical aspects.

References

1. Wood RE, Boat TE, Doershuk CF (1976) Cystic fibrosis. Am Rev Respir Dis; 113: 833–877
2. Shwachman H, Kulczycki L (1958) Long term study of one hundred five patients with cystic fibrosis. Am J Dis Child 96: 6–15
3. Shwachman H (1960) Therapy of cystic fibrosis of the pancreas. Pediatrics 25: 155–163
4. Shwachman H, Kulczycki L, Khaw KT (1965) A report on 65 patients over 17 years of age, studies in cystic fibrosis. Pediatrics 36: 689–699
5. Shwachman H, Holsclaw D (1969) Complications of cystic fibrosis. N Engl J Med 281: 500–501
6. Cooperman E, Park M, McKee J, Assad J (1971) A simplified cystic fibrosis scoring system. Can Med Assoc J 105: 580–586
7. Chrispin A, Norman A (1974) The systematic evaluation of the chest radiograph in cystic fibrosis. Pediatr Radiol 2: 101–105
8. Matthews D, Warner O, Chrispin A, Norman A (1977) The relationship between chest radiographic scores and respiratory function tests in children with cystic fibrosis. Pediatr Radiol 5: 198–200

Date of final acceptance: November 1981

Academisch Ziekenhuis Rotterdam
Sophia Children's Hospital
Gordelweg 160
Post box 70029
NL-3000 LL Rotterdam
The Netherlands