Bronchiectasis in Children*

ALBERTO CHATTAS, M.D., F.C.C.P.
Cordoba, Argentina

Since the classic description of the bronchiectasis by Laennec in 1819, our knowledge about dilatation of the bronchial tree has greatly increased through:

1) Anatomopathological studies.
2) Bronchographic studies.
3) Endoscopy.
4) Surgical treatment.

Modern bronchographic technic has made easy the early discovery of bronchiectasis. It is also applicable routinely during the first years of life, permitting modern surgical treatment, with very little risk and excellent results even in extensive bilateral cases (Overholt and Langer). Unfortunately, we still too frequently see children harboring bronchiectasis for many years without diagnosis and dying of some broncopulmonary complication.

Reviewing the official mortality statistics in the city of Córdoba, we see that from 1930 to 1940 no case of death caused by bronchiectasis in children under 15 years of age, was reported. No case was reported under the label of chronic bronchitis or nonspecified bronchitis in 1930. In 1935, 4 cases were reported, all of them under 9 years of age (2 chronic and 2 nonspecified); in 1940, 3 cases were reported, all of them under 1 year of age (1 chronic and 2 nonspecified). This lack of figures relative to mortality due to bronchiectasis in children shows obviously, how the diagnosis of this disease in the medical practice is overlooked, even now.

In our city, the third largest in the Argentine, with a population of 325,000 inhabitants, its infant mortality due to pulmonary disease is: 19 per cent of the general mortality from 0-1 year of age; 20.4 per cent of the general mortality from 1-4 years of age; 8.8 per cent of the general mortality from 5-9 years of age.

In the Children's Hospital of Córdoba during the last 10 years, of 27,636 patients, only 60 cases were hospitalized on account of bronchiectasis, that is 0.2 per cent.

It is estimated that in children, bronchiectasis represents about 5 per cent of all diseases. Bonniot,2 reports that one third of the

children with bronchiectasis die in 2-3 years after acquiring the disease. Head states that persons who have developed bronchiectasis during childhood live only to 40 years of age. Perry and King, studying 96 patients with bronchiectasis in the first decade of life, noted that 65 per cent die within 20 years and 90 per cent within 30 years.

**Bronchiectasis in Pediatrics**

The study of the disease in adults shows the frequency of early symptoms during childhood, an age in which a number of illnesses complicate bronchial changes (especially whooping cough). Laennec said that out of every four bronchiectasis in the adult, three had their origin in childhood. According to other reports, symptoms of bronchiectasis in adults is traceable to the first decade of life in 27 per cent and to the first two decades in 57 per cent.

In the first decades of life, the symptoms of bronchiectasis are intermittent, and often masquerading as acute lung diseases, especially pneumonia. We always remember the old teaching of Czerny who said, "that every child that has repeated acute pneumonia with the same localization of the lesion, should make the physician think of bronchiectasis."

The responsibility of the pediatrician is greater than that of the general practitioner, because he has the primary responsibility in the early recognition of the disease and stands in "the first line of defense" as Brennemann said. The diagnosis can be frequently suspected from the history of the patient and from the physical examination; however a precise diagnosis can only be made as we mentioned before, by bronchographic studies.

**Classification of Bronchiectasis in Children**

Bronchiectasis is classified as congenital and acquired. The congenital type was considered till not many years ago, as the most frequent, but new systematic investigation of the bronchial tree shows now that acquired bronchial changes are the most common during the first decade of life. Congenital bronchiectasis is produced by a lack of development in the embryonic stage that Castex calls "alveolo-agenesis-bronchiectasis."

Recent researches attribute these congenital malformations to virus diseases, acquired by the mother during the first 3 months of pregnancy (Erickson, Conte and his associates). According to the experimental work of Warkany, the malformation in the embryo may be due to a lack of vitamin A or B. Kartagener published his observations on bronchiectasis in "Situs viscerum inversus" and describe what is known as the "triad of Karta-
gener" that is, the coexistence of bronchiectasis, sinusitis and congenital dextrocardia.

**Pathogenesis of Bronchiectasis in Children**

Acquired bronchiectasis may result from the following diseases:
1) Bronchial infections, especially whooping cough. 2) Measles.
3) Chronic pneumonitis, sclerosis and lung abscesses. 4) Foreign bodies. 5) Atelectasis. 6) Tuberculosis. 7) Infection of tonsils and paranasal sinuses. 8) Allergic catarrhs.

Especially in children under 5 years of age, after acute or subacute bronchitis, there is a change in the bronchial mucous membrane, with a thickening of it. These changes are also observed after whooping cough, measles and repeated grippes. Following these changes of the mucous membrane, alterations in the basal stratum of the bronchial wall appear, becoming chronic and developing into definitive bronchiectasis. Transitory bronchial enlargement during whooping cough may be due to muscular paralysis of the bronchus in the neighboring zones of the inflamed mucosa. We must remember that histologically, the small bronchi are principally formed by muscles that occupy the greatest part of the bronchial wall and contribute with their tension to the preservation of the normal bronchial form.

Roentgenologic studies of the chest are of great value in discovering atelectasis in some segments of the lung. Atelectasis sometimes preceeds, coexists or follows a bronchiectasis and it may be due to bronchial obstruction produced by infections (Ansprech).

It is important to bear in mind that bronchiectasis involving an entire lobe may be produced by foreign bodies. We shall briefly describe the following cases:

A girl, 11 years of age, came to us with a bronchopulmonary disease which had begun 5 years before. During the later months, she had repeated hemoptyses. She came to the Children's Dispensary of the Instituto de Tisiología de Córdoba (Director Prof. G. Sayago) because her private physician thought of tuberculosis. X-ray film showed a shadow in the right cardiophragnostic angle.

We ruled out tuberculosis because the tuberculin reaction was negative. A lateral x-ray film showed very clearly the presence of a screw in the basal segment of the right lower lobe. Neither the mother nor the child knew of the presence of a screw in the lung. After a careful inquiry, they both remembered that 5 years before, while the girl had whooping cough, she played with a box of screws and nails. After two bronchoscopic sessions, the foreign body was removed: it was a rusty screw, one inch long (Figs. 1 and 2).

Hemoptyses stopped, but the cough persisted. A bronchogram was performed, showing very clearly bronchiectatic changes in the pulmonary segment obstructed by the screw.
Mycotic infection as a primary disease in the lung may also produce bronchiectasis:

We had such a case: a child, 5 years of age, came to the Children's Hospital (Prof. J. M. Valdes) in very poor condition. She had fever, cough and expectoration for eight months. An x-ray film showed almost complete atelectasis of the entire right lung. Bronchoscopy revealed abundant secretions coming from the right stem bronchus which was narrowed; unfortunately due to his poor condition bronchogram was not done. The child died soon after admission. Post mortem studies revealed typical fungus colony, with bronchiectasis and multiple abscesses (Fig. 4).

Bronchiectasis and Atelectasis in Children

Huizinga\textsuperscript{13} of Holland, reporting on 40 cases of bronchiectasis in children, emphasized the importance of atelectasis in producing bronchial alterations. Clinical examinations reveal evidence of a condensed lung. Fluoroscopic or roentgenographic studies will show a triangular shadow, corresponding to an atelectatic segment. Anspach, Ellis, Brennemann, Holinger\textsuperscript{6,12,14,15} and others have published several papers dealing with the relationship between bronchiectasis, collapsed lung (shrunken lung) and the triangular shadow seen on the x-ray film. Anspach\textsuperscript{12} examining thousands of children's x-ray pictures of the chest found nearly 1 per cent of triangular shadows.

We wish to call attention to the seriousness of atelectasis, especially during the first two years of life, not only because of possible bronchiectasis but also because of secondary infections that may occur. Sometimes the new born suffers from atelectasis, which is not noticeable and which may be the cause of a partly shrunken lung. During the first year of life, thickening of bron-
chal secretions may sometimes cause obstruction of the small bronchi. This obstruction of the air passage reduces the ingress of air until it causes atelectasis.

We feel that all the broncopulmonary diseases should be followed very carefully by x-ray studies, in different projections. Many small opaque triangular areas can be easily overlooked when the film is taken only in one projection. We pediatricians are responsible to a certain extent for the development of bronchiectasis if we fail to make an accurate diagnosis.

In our practice we have found certain simple measures valuable in preventing or curing atelectasis. We resort to medication that liquify the bronchial secretion; postural drainage, frequent changes of position, naso-tracheal aspiration; and as a last resort bronchoscopic aspiration. In new borns we find that the results obtained by the use of carbogen (95 per cent O₂ + 5 per cent CO₂) in aerating the lung are very gratifying.

We shall now show the outcome of two cases of atelectasis developed in early life, not diagnosed until complications developed:

A girl, 9 years old, had whooping cough when five years of age and measles at the age of six. For the last 7 years she complained of productive cough. X-ray picture showed a basal triangular shadow beneath the cardiac area (Fig. 5). Bronchographic studies were not done at that time. Operative specimen revealed a small fibrotic and shrunken lower lobe with bronchiectatic changes (Children's Hospital, Prof. J. M. Valdes).

A girl, 6 years of age, had bronchitis when 6 months old and at three years of age had whooping cough. Plain x-ray picture (taken when she came to the Hospital, Surgical Department, Prof. Dr. J. M. Allende), showed an opaque zone in the lower half of right lung with a marked displacement of the heart towards the affected side (Fig. 6). The bronchogram revealed saccular bronchiectasis in the right upper lobe (Fig. 7). The parents refused surgical intervention.

SUMMARY AND CONCLUSION

1) Atelectasis is frequent in childhood and must be considered as an important cause of bronchiectasis.

2) It is the duty of the pediatrician to resort to all the diagnostic means at our disposal to make an early diagnosis.

3) Bronchiectasis in the adult is a process which frequently originates in the first or second year of infancy.

4) Bronchial changes can take place in whooping cough and measles. These changes may not be permanent, and in this prebronchiectatic stage, medical treatment can be sometimes successful.

5) If medical treatment fails, bronchiectasis becomes defin-
ately established and irreversible; only by surgical treatment can a permanent cure be obtained.

6) Surgical treatment must not be postponed. Children tolerate the intervention better than adults.

7) Our experience deals with 10 cases, 4 boys and 6 girls; between 4 and 13 years of age; 7 were operated upon, and 3 refused operation. Of the 7 operated cases, bronchiectasis was present in 4 in the left lower lobe and in 3 in the right lower lobe. They all made complete recovery; one of the nonoperated patients died. One of the cases showed the triad of Kartagener. In all cases the bronchiectasis was localized in the lower lobe, with exception of one that had bronchiectasis in the right upper lobe, and in one, the bronchiectasis of the left lower lobe was associated with bronchiectasis of the lingula.

RESUMEN Y CONCLUSIÓN

1) La atelectasia es frecuente en la niñez y debe ser considerada como una causa importante de bronquiectasia.

2) El especialista en pediatría tiene el deber de valerse de todos los medios de diagnosticar a su disposición a fin de hacer un diagnóstico temprano.

3) La bronquiectasia en el adulto es un proceso que frecuentemente se origina en el primero o segundo año de la infancia.

4) En la tos ferina y la alfombrilla pueden ocurrir alteraciones bronquiales que no sean permanentes y en esta etapa pre-bronquiectásica el tratamiento médico es a veces eficaz.

5) Si fracasa el tratamiento médico, la bronquiectasia queda establecida definitivamente y es irreversible, solamente mediante el tratamiento quirúrgico puede obtenerse una curación permanente.

6) No debe aplazarse el tratamiento quirúrgico. Los niños soportan la intervención mejor que los adultos.

7) Nuestra experiencia incluye 10 casos: 4 niños y 6 niñas, de 4 a 13 años de edad. Se operó a 7, tres rehusaron la operación. De los 7 casos operados la bronquiectasia afectaba el lóbulo inferior izquierdo en 4, y el lóbulo inferior derecho en 3. Todos los operados se repusieron por completo, mientras que uno de los pacientes no operados murió. Uno de los casos manifestó la terna de Kartagener. En todos los casos la bronquiectasia estaba localizada en el lóbulo inferior, con la excepción de uno que tenía bronquiectasia en el lóbulo superior derecho y otro en el que la bronquiectasia del lóbulo inferior izquierdo estaba asociada con bronquiectasia del proceso lingual.
REFERENCES