RAPIDLY expanding congenital cysts of the lung may appear suddenly during infancy. Recognition is simple and prompt surgical treatment is effective. The condition is not common enough to be at one's diagnostic fingertips but is usually so characteristic and dramatic that one case leaves a lasting impression. Six congenital lung cysts, all emergencies, and all in infants, were seen within two years and form the basis of this report.

There are also symptomless congenital lung cysts of relatively small size which come to light when, for some reason or other, a roentgenogram of the chest is made. Advice concerning removal of such cysts is slanted by the past experience of the attending staff. Cysts, also called pneumatoceles or loculated emphysema, which develop as a sequel to staphlococcic pneumonia are the most common cause of radiolucent shadows in the lungs of infants. These cysts and small congenital cysts are those for which Caffey1 in a recent report advises conservative treatment. Advice concerning such cysts obviously will be based on many considerations not pertinent to this discussion.

Interest is focused here only on the congenital lung cysts which are large enough to cause severe symptoms and demand prompt treatment. Acquired lung cysts following staphylococcic pneumonia during infancy may become emergencies because of size or rupture, but are not included in this review.

SYMPTOMS AND DIAGNOSIS

Cough, anorexia and moderately rapid breathing may be noted as early symptoms but are not impressive. Only when over a period of days, or more often hours, severe dyspnea and cyanosis are noted may the diagnosis be suspected. Physical examination of such a child in respiratory distress is, of course, in order but not too satisfactory. One will find diminished breath sounds and hyperresonance on the affected side with the mediastinal structures pushed over to the opposite side. If the child is in real respiratory distress, more will be accomplished by placing it in oxygen and immediately preparing for roentgenographic examination, than by going through the labor of an academically satisfactory physical examination.

Presumably the distressed infant will be seen in the nursery. The child should be placed in a covered crib or isolette, with a high concentration of oxygen, and should be sent to the x-ray department. If such conveniences are not at hand, the child may be transported in a buggy or on a cart while giving oxygen from a portable tank with a funnel placed over the child's face. Bedside RGs of an infant breathing 60 to 75 times a minute are often nothing more than a mass of misleading fuzzy shadows. With all in readiness, the infant can be taken out of the oxygen safely for the few moments required for making the RGs with a fast machine. The infant should be kept in the upright position with the hands above the head while the antero-posterior and lateral views are made. The infant

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Fig. 1. Case 1, R.K. Admission P-A RG showing marked mediastinal shift to left due to cysts in right lung.

Fig. 2A and B. Case 2, J.H. Preoperative P-A and lateral RGs showing location of cysts in right lung. Note marked mediastinal displacement to left.
is quickly replaced in oxygen but is kept in the department until the films have been
developed and proven satisfactory. This rather detailed review of the simple procedure
of making a roentgenographic study of a dyspnoic infant is justified only because good
RGs are essential for the diagnosis and differentiation of translucent shadows in the chest
of the newborn infant. Five possible diagnoses must be considered as the RGs are studied:
congenital lung cysts, congenital diaphragmatic hernia, pneumatoceles secondary to
staphylococcic pneumonia, tension pneumothorax and congenital lobar emphysema. The
outstanding symptoms—dyspnea and cyanosis—are the same in each condition.

Congenital lung cysts, fortunately, are confined usually to one lobe or a portion of one
lobe. They may be single but usually are loculated. The RG shows the clear air spaces
(Figs. 1 and 2) divided by septa into large segments. Mediastinal structures are pushed
to the opposite side and the adjacent lobe or lobes are collapsed and usually visible as
triangular shadows.

A congenital diaphragmatic hernia on the left side is usually recognizable in a new-
born child by the characteristic appearance of loops of bowel in the left hemithorax and
by the displacement of the mediastinal structures to the right. However, if the diaphrag-
matic defect is on the right side and only a few loops of distended bowel are in the chest,
it may be difficult to make a definitive diagnosis without giving the infant some contrast
media by mouth and watching its progress through the bowel.

Pneumatoceles usually do not present a diagnostic problem if evidences of the etiologic
pneumonia are still present. The history of fever, the clinical and roentgenographic find-
ings of co-existing pneumonia practically always clinch the diagnosis of pneumatoceles.
Aspiration of fluid from the chest and recovery of staphylococci confirm the diagnosis of
pneumatoceles following pneumonia.2 It is possible to mistake pneumatoceles for dia-
aphragmatic hernia.

Tension pneumothorax may be spontaneous in the newborn infant or may be the
result of a ruptured pneumatocele. In the former condition, half the chest is filled with
air and the collapsed lung as an oval shadow is visible at the hilus. In the latter condi-
tion the lung may be only partially collapsed and will most likely still show signs of
pneumonia. Free air will be seen in the chest and usually an unruptured cyst or cysts will
be seen in the uncollapsed portion of the lung.

Congenital lobar emphysema is a relatively infrequent condition which has not often
been described.8 One lobe of the lung, generally the upper lobe on either side, is greatly
distended, pushing the mediastinal structure to the opposite side and collapsing the
uninvolved lower lobe. It is very difficult and sometimes impossible to differentiate this
condition from a lung cyst. The only clue is the finer lung markings in the former condi-
tion. If an attempt is made to relieve the cyanotic infant by aspiration and no air can be
withdrawn one promptly becomes suspicious that one is dealing with an emphysematous
lung rather than a lung cyst.

Two illustrative cases are briefly outlined:

Case 1, R.K.: A 5 day old male infant was admitted to the hospital by ambulance with a history
of rapid respiration and cyanosis of 24 hr. duration. His birth had been at full term, breathing
spontaneous and normal. On the fourth day of life cyanosis was seen after a feeding. Chest RGs
revealed large cysts in the right lung (Fig. 1). It was impossible to be sure which lobe or lobes were
involved.

The infant was afebrile and slightly cyanotic. Respiration rate was 60/min. Right hemithorax
was hyperresonant and breath sounds were absent. Heart tones were heard far to the left of normal
Because dyspnea and cyanosis increased during a few hours of observation, surgery under intratracheal anaesthesia was performed as an emergency the day of admission. Chest was opened through the right 5th interspace and the right lower lobe containing a number of cysts was resected. Upper and middle lobes, completely collapsed, each about the size of a pecan nut, could be partially expanded by pressure on the anaesthetic bag. Chest was drained and closed snugly in layers. When the child had been returned to its crib, the end of the drainage tube was placed under water in a bottle to form a seal.

Pathologic examination confirmed the diagnosis of lung cyst.

Convalescence was uneventful and the child was discharged 2 wk. after operation, at which time the right middle and upper lobes had expanded sufficiently to fill the right thorax.

Case 2, J.H.: A 1 mo. old male infant was admitted by ambulance to the hospital with severe dyspnea and marked cyanosis. Fortunately, roentgen films taken at another hospital and accompanying the child were diagnostic of a lung cyst (Fig. 2). The baby was given emergency oxygen in the admitting room and the chest was quickly aspirated. The infant improved promptly.

A history was then obtained. This vigorous, apparently normal 4.1 kg. infant had been delivered spontaneously. The child was taken home and continued healthy until 2½ wk. of age, when it had an attack of dyspnea and vomiting. A change of formula stopped the vomiting. Dyspnea continued and cyanosis appeared. The infant was returned to the hospital where it had been born and roentgenographic studies of the chest were made. On 2 occasions the chest was aspirated and each time the infant was improved for a few days. Rather suddenly the symptoms became severe and the patient was transferred for emergency treatment.

After the chest had been aspirated in the admitting room, preparations were made for immediate surgery. A 20 cc. syringe and needle were kept at the baby's side and a resident remained in constant attendance.

The anaesthesiologist, Dr. William O. McQuiston, intubated the infant but could not obtain even first plane anaesthesia because the exchange of gases was so inadequate. The chest was aspirated again but cyanosis persisted. It was obvious that immediate relief was necessary. The skin of the right chest was hastily prepared and without towels, drapes or gowns, and with no attention to hemostasis the right chest was opened. The tense cyst in the right upper lobe herniated out between the ribs separated by finger traction. Immediately the heart, which had practically stopped, began to beat normally, the cyanosis disappeared, the child could now be anaesthesized and the operation could be continued in an orderly fashion. It was found that the cystic disease was confined to the anterior apical segment of the right upper lobe. This segment was resected. The chest was drained, closed, and the infant treated as reported in Case 1. The pathologist confirmed the diagnosis of lung cyst.

Convalescence was uneventful and the child was discharged from the hospital on the eleventh day following surgery.

Treatment

This paper is not concerned with the nonemergency type of lung cyst which can be studied leisurely and treated as circumstances warrant. Too little attention has been given the infant who, because of an expanding lung cyst, becomes visibly cyanotic and distressingly dyspnoeic. In fact, it is only 12 years since Fischer et al.4 reported the first successful emergency resection of a lung cyst in a one month old baby.

So soon as a definitive diagnosis of an expanding lung cyst has been made and confirmed by roentgenographic examination, the infant is literally followed by a physician with a large syringe and needle in his hands while preparations are being made for surgery. Aspiration of air from the cyst should be done only to relieve increasing dyspnea and deepening cyanosis; it is never done as an elective procedure. If aspiration has become necessary to relieve symptoms, the child from then on must be watched still more carefully because a pneumothorax may have resulted from puncturing the cyst. One may
be sure the cyst will promptly refill with air. Attaching continuous suction to a needle in the chest leads only to trouble. If the child for some reason must be transported to another institution it is probably safest to keep the child in oxygen and be ready to aspirate the chest as often as necessary until surgery can be done. (See Case 2.)

Surgical treatment presents no difficulties if an anaesthesiologist is available who can gently and surely intubate the infant and maintain a proper exchange of gases.

The chest is opened through a properly placed intercostal incision and the greatly expanded cyst is allowed to herniate itself out of the wound. So soon as the cyst has extruded itself from the chest, the infant will breathe better and the surgeon can unhurriedly proceed with resection of a segment or a lobe of lung, as circumstances demand. After resection is completed, the chest is drained with a catheter which is attached to a water seal bottle after the infant is returned to its crib.

Postoperative care consists merely of keeping the infant in oxygen for 24 hours, more or less, giving the proper amount of antibiotics and providing adequate humidification if there is any sign of postanaesthetic laryngotracheitis.

SUMMARY

The diagnosis and treatment of congenital lung cysts producing severe symptoms are discussed. The cases of six infants form the basis of this report. All had such severe dyspnea and cyanosis that emergency operations were necessary. All six infants made uneventful recoveries. The youngest child was five days old.

REFERENCES


SPANISH ABSTRACT

Quistes Congénitos del Pulmón en la Infancia

Aun cuando los quistes congénitos del pulmón no son tan frecuentes como para pensar en ellos a cada momento, en ocasiones presentan síntomas tan severos y drásticos que los casos se convierten en problemas de emergencia. En este artículo se estudian seis casos que requirieron tratamiento inmediato quirúrgico. Los síntomas iniciales son tos, anorexia y polipnea, seguidas en unas cuantas horas o días de disnea intensa y cianosis; clínicamente se pueden confundir con hernia diaphragmática congénita, neumatocele secundario a neumonía estafilocócica, neumotórax a presión y enfisema lobar congénito, pues los signos más notables (disnea y cianosis) son los mismos en todos ellos; el diagnóstico diferencial se hace por medio de los rayos X. El tratamiento es fundamentalmente quirúrgico al presentarse la emergencia: aspiración de aire de los quistes para aliviar al niño de la disnea y la cianosis, e inmediatamente después toracotomía con resección segmentaria o lobar del pulmón, según las circunstancias. El postoperatorio consiste en la administración de oxígeno, antibióticos y vapor.

Los seis casos presentados se recuperaron favorablemente. El más pequeño tenía cinco días de nacido.
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