

## MORPHOLOGIC ANALYSIS OF FOREGUT CYSTS IN THE THORAX\*

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A SYSTEM of morphologic analysis currently under development at the Armed Forces Institute of Pathology was applied to the analysis of 86 congenital thoracic cysts of foregut origin after application of rigorous histologic review and classification.

The roentgenograms of the 86 cases were compared using the following basic qualitative and quantitative roentgenologic parameters: density; border; shape; size; location; associated bone abnormalities; and frequency of communication.

### MORPHOLOGIC ANALYSIS

#### DENSITY (Table I)

Most of the cysts in the series were solid and of homogeneous water density (Fig. 1A). Air was contained in only 4 respiratory cysts and 1 enteric cyst and a definite air-fluid level was noted in 3 of the respiratory cysts and the enteric cyst (Fig. 1, B and C). No calcification was ever seen within a cyst but a peripheral type of calcification occasionally occurred in both respiratory and enteric cysts (Fig. 2A). Histologically, the latter type of calcification was observed to be in dystrophic tissue in areas of inflammatory residual (Fig. 2B).

#### BORDERS

Where the borders of foregut cysts were observable they were sharply delineated (Fig. 3A). In the majority of these cysts however, it was not possible to evaluate their borders completely since they often were located so deeply within the mediasti-

num that they were obscured in varying degrees (Fig. 3, B and C).

#### SHAPE (Table I)

The shape which was encountered in most of the respiratory cysts and all of the enteric cysts was categorized as spheroid (round to oval) (Fig. 4, A and B). A lobulated appearance was observed in 8 of the respiratory cysts (Fig. 4, C and D).

#### NUMBER

All of the foregut cysts were single. The cases first considered as multiple were later histologically rejected as probably being the result of infection rather than of errors in embryogenesis.

#### SIZE (Table II)

A relative evaluation of the size of these cysts was made by comparison with the posterior rib interspaces: small=1 interspace; medium=1-2; large=2-3; and extra-large=over 3. There were only 3 respiratory cysts in the small category. Most of the respiratory cysts were in the medium and large ranges, while the enteric cysts tended to fall into the extra-large groups (Fig. 5, A-D).

#### LOCATION (Table III)

A mediastinal location accounts for the majority of the respiratory cysts and all of the enteric cysts. There was a right-sided predominance in both groups (Fig. 1A). In a superior-inferior orientation (considering the level of the carina as the dividing

\* Presented at the Seventy-fourth Annual Meeting of the American Roentgen Ray Society, Montreal, P.Q., Canada, September 25-28, 1973.

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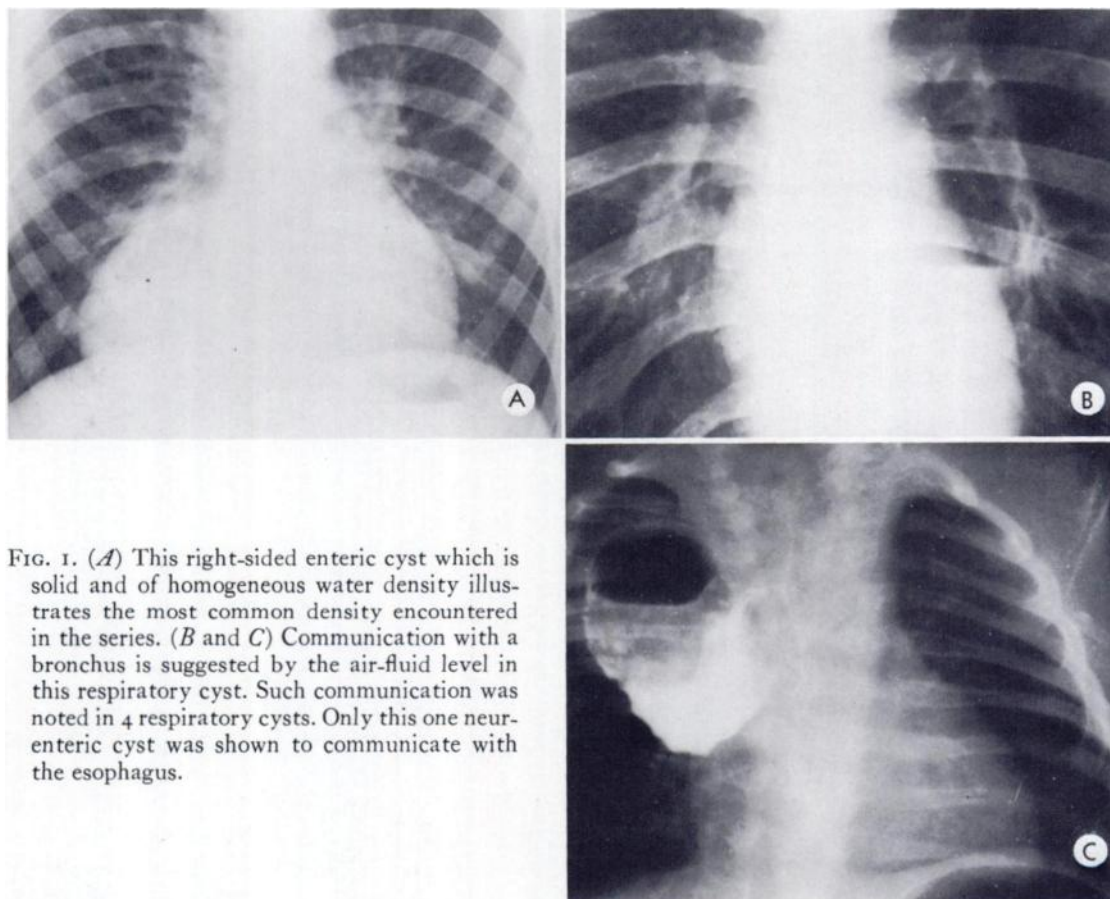


FIG. 1. (A) This right-sided enteric cyst which is solid and of homogeneous water density illustrates the most common density encountered in the series. (B and C) Communication with a bronchus is suggested by the air-fluid level in this respiratory cyst. Such communication was noted in 4 respiratory cysts. Only this one neuroenteric cyst was shown to communicate with the esophagus.

TABLE I  
FOREGUT CYSTS IN THE THORAX:  
DENSITY AND SHAPE

	Respiratory (77)		Enteric (9)	
	No.	Per Cent	No.	Per Cent
<i>Density</i>				
1. Water	73	94.8	8	88.9
2. Air (communicating)	4	5.2	1	11.1
3. Calcium (peripheral)	2	2.6	1	11.1
<i>Shape</i>				
1. Spheroid	69	89.6	9	100
2. Lobulated	8	10.4		

TABLE II  
FOREGUT CYSTS IN THE THORAX: SIZE

	Respiratory (77)		Enteric (9)	
	No.	Per Cent	No.	Per Cent
<i>Size</i>				
Small = 1 interspace	3	3.8	0	
Medium = 1-2 inter- spaces	35	45.6	2	22
Large = 2-3 interspaces	31	40	1	11
Extra-large = over 3	8	10	6	66

point), respiratory cysts were more common in an inferior location, while the enteric cysts tended to be more superior.

The respiratory cysts tended to be juxtaposed about the carina and therefore most were middle mediastinal (Table IV). Of the carina-based group, 15 of 30 extended posteriorly, 13 inferiorly, and only 2 anteriorly (Fig. 6, A-C). Only a few of the respiratory cysts were found posteriorly at some distance from the carina (Fig. 7A), while almost all of the enteric cysts were in such a posterior position.

A number of the mediastinal cysts were

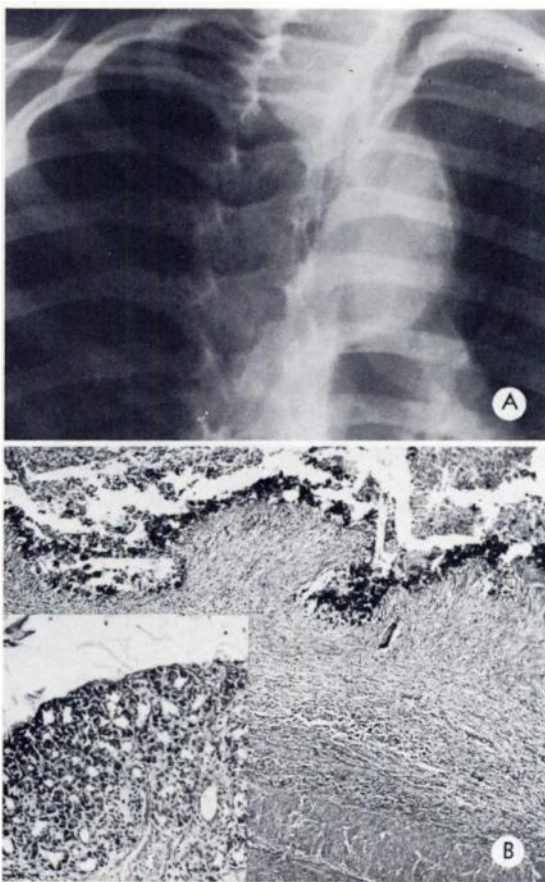


FIG. 2. (A) This enteric cyst reveals the type of peripheral calcification that occurred in 2 respiratory cysts and 1 enteric cyst. (B) In a histologic field from the cyst in A, the epithelium is denuded, and a layer of calcium salts lines the chronically inflamed wall. (Insert) Layers of smooth muscle and a focus of gastric glands identify the cyst as enteric.

TABLE III  
FOREGUT CYSTS IN THE THORAX: LOCATION

	Respiratory (77)		Enteric (9)	
	No.	Per Cent	No.	Per Cent
<i>Mediastinal</i>	66	86	9	100
Right	44	68	6	66
Left	19	29	3	33
Midline	2	3		
Indeterminate	1			
Middle	58	87	1	11
Posterior	8	12	8	88.9
Superior	21	32	6	66
Inferior	45	68	3	33
<i>Intrapulmonary</i>	11	14	0	
Right	6	54.5		
Left	5	45.5		

demonstrated to have a close relationship with the esophagus and 8 cases were actually identified within the wall of the esophagus (Fig. 7B). This raises the question of classification of esophageal cysts which will be discussed in the following section on pathologic review. One of the group contained cartilage and was therefore identified as respiratory in origin, but the other 7 cases were less differentiated and should probably be classified as undifferentiated foregut cysts (see the following section).

Only 11 of the respiratory cysts were

TABLE IV  
MIDDLE MEDIASTINAL RESPIRATORY CYSTS (58)

	No.	Per Cent
Carina-based	30	51.5
Posterior	15	
Inferior	13	
Anterior	2	
Superior (paratracheal)	11	19
Esophageal wall	8	13.8
Inferior (retrocardiac)	5	8.6
Indeterminate	4	6.9

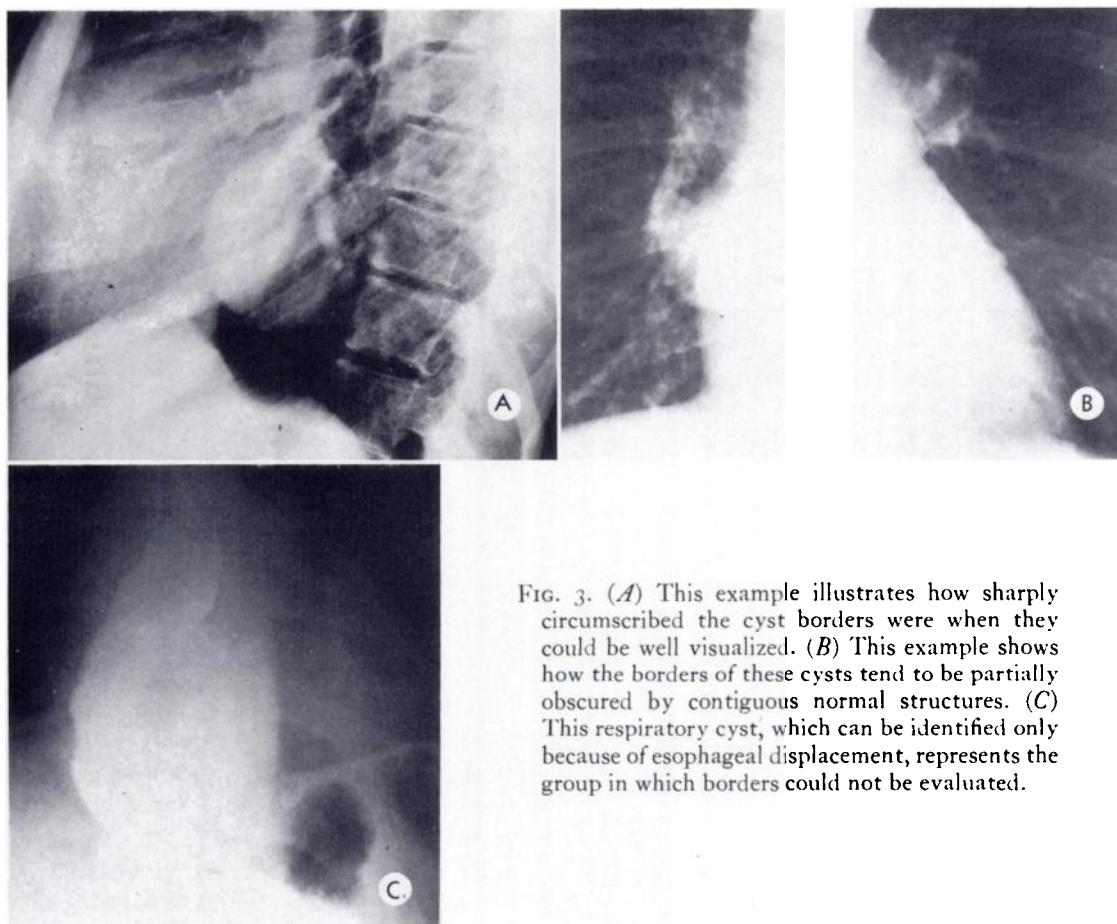


FIG. 3. (A) This example illustrates how sharply circumscribed the cyst borders were when they could be well visualized. (B) This example shows how the borders of these cysts tend to be partially obscured by contiguous normal structures. (C) This respiratory cyst, which can be identified only because of esophageal displacement, represents the group in which borders could not be evaluated.

*intrapulmonary* (Fig. 7C). This group differed from the mediastinal cysts by having a nearly equal right-left distribution, and more importantly included all respiratory cysts that communicated and contained air or air-fluid levels. A lobulated shape was observed in 3 of this pulmonary group.

#### ASSOCIATED ANOMALIES

Associated vertebral body *anomalies* were observed only in the enteric cysts. Six of the 9 enteric cysts were thus properly identified as *neurenteric* cysts, since there were no histologically distinguishing features for separating the neurenteric and enteric cysts. These anomalies included hemivertebrae, butterfly vertebrae, and scoliosis. There was 1 case with a cervical spine anomaly, and it should be emphasized

that the spinal anomaly may be located above the level of the cyst itself (Fig. 8).

#### REVIEW AND CLASSIFICATION OF PATHOLOGIC MATERIAL

The microslides on each case were reviewed. The cyst was first judged to be either intrapulmonary or mediastinal, on the basis of associated pulmonary alveolar parenchyma. However, if the lesion had pleural encapsulation and showed dilatation of bronchioles and alveoli, the possibility of extralobar sequestration of the mediastinum was considered. Cysts lined by cuboidal epithelium or without epithelium and having no features suggestive of respiratory or enteric origin were excluded from the study. Those cysts having a mucus-secreting columnar epithelium

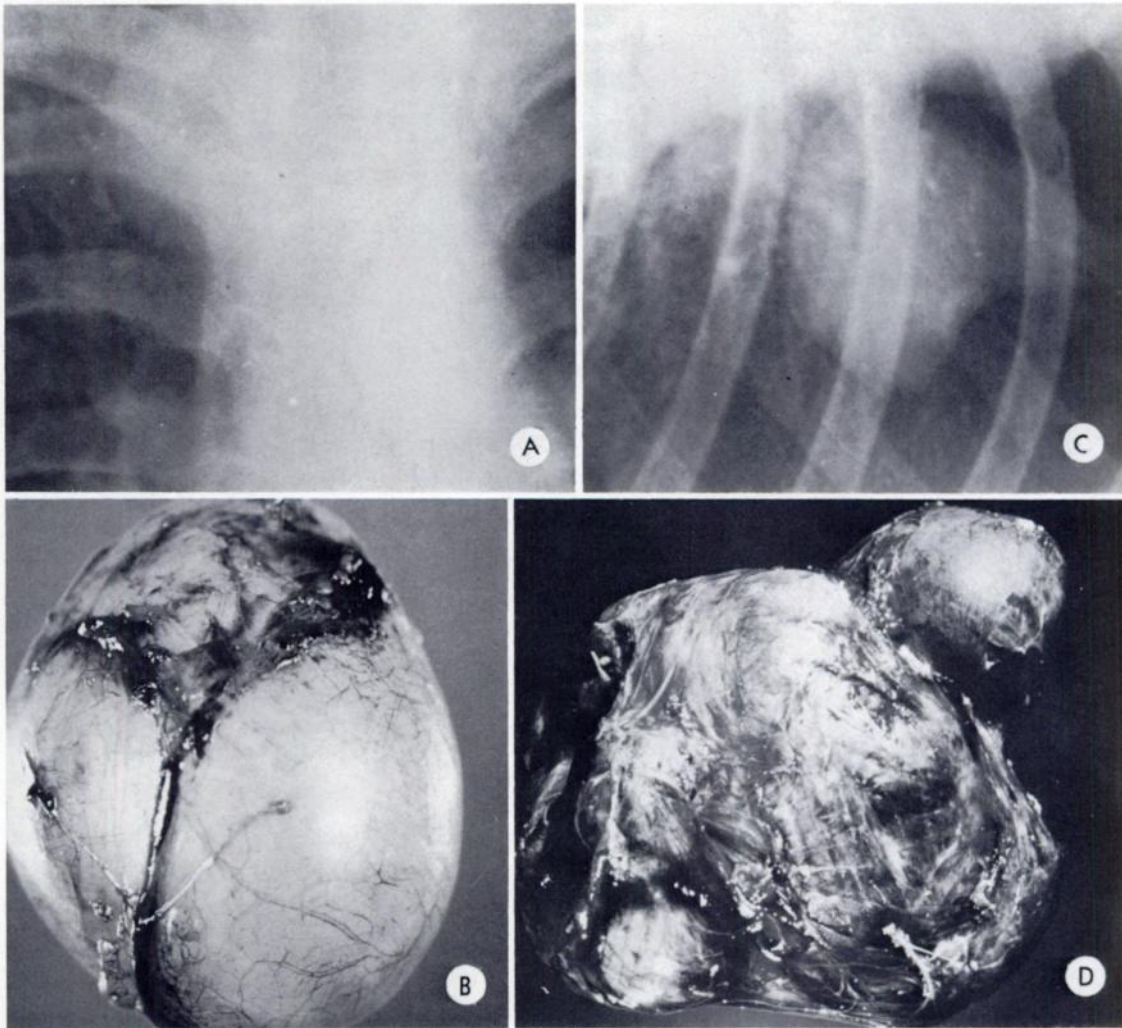


FIG. 4. (A) The most common shape is illustrated by this respiratory cyst and is categorized as spheroid. This shape was found in 69 respiratory cysts and all of the enteric cysts. (B) The gross specimen of this respiratory cyst correlates with the roentgenographic appearance of a spheroid (round to oval) mass. (C) This lobulated respiratory cyst is a striking example of a polycyclic or lobulated mass. This lobulated appearance was observed in 8 respiratory cysts, but was not seen in the enteric group. (D) This gross specimen of another histologically confirmed respiratory cyst would result in a similar roentgenographic pattern of a lobulated mass.

were considered to be of enteric origin and the presence of gastric glands, parietal cells, layers of smooth muscle, and associated areas of esophageal (squamous) or small intestinal epithelium was sought. The presence of chronic inflammation, ulceration, and calcification was also noted. Those cysts with a squamous or respiratory epithelium were further analyzed in terms of the presence of seromucinous glands of

the bronchial type, mucous glands, plates of cartilage, layers of smooth muscle, skeletal muscle, and associated inflammation, ulceration, and ossification of the cartilage. The microscopic findings were then compared with the available clinical data and previous pathologic interpretation.

Among the intrapulmonary cysts, those cysts without respiratory epithelium were

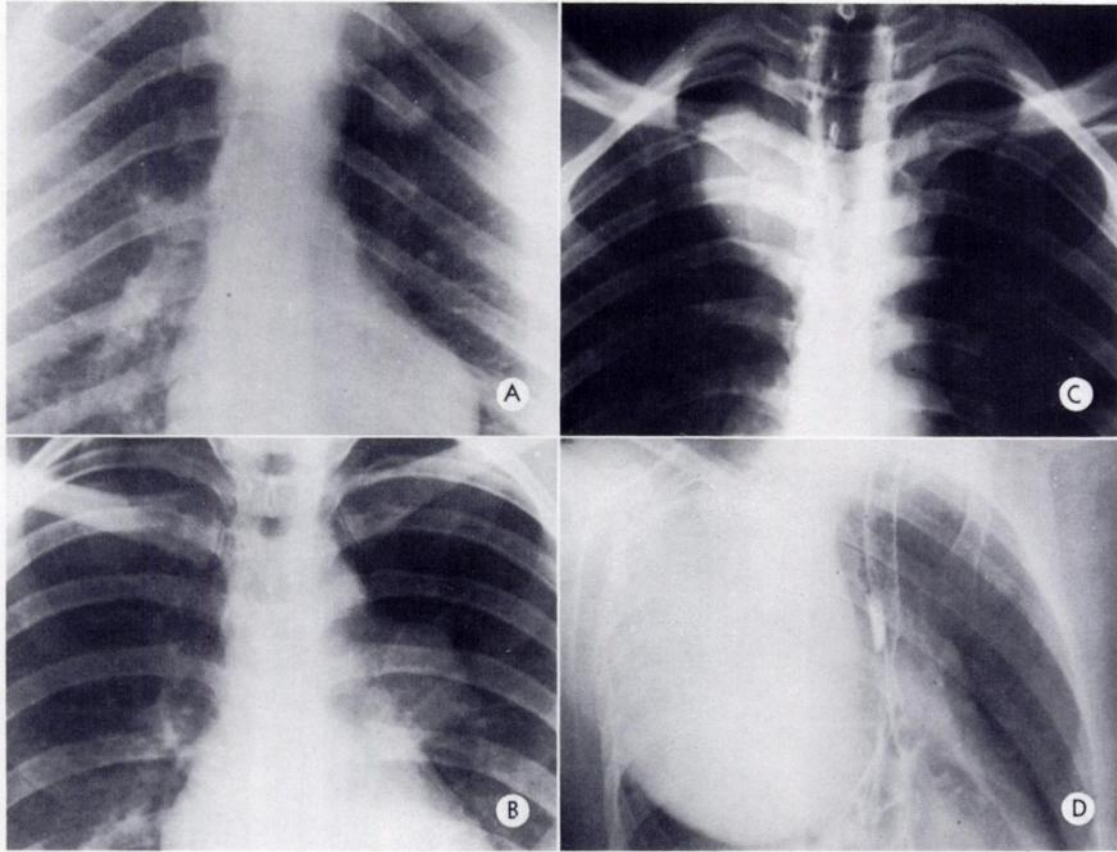


FIG. 5. (A-C) These 3 respiratory cysts demonstrate the smallest cyst in the series (A) and the most common size range of medium to large (B and C). (D) This neurenteric cyst represents the extra-large group. There were 6 of 9 enteric cysts and only 8 of 77 respiratory cysts in the extra-large range.

discarded. The presence of acute or chronic inflammation in the cyst wall raised the possibility of a postinfectious origin for the cyst. Although such a finding might represent infection of a congenital cyst, these cases were excluded from the study in most instances. The association of an inflamed cyst with areas of bronchiolectasis, acute and chronic bronchiolitis and interstitial fibrosis of the parenchyma adjacent to the cyst was taken as evidence of a probable postinflammatory state and these cases were all excluded from the study. If the cyst was associated with a rather uniform dilatation of adjacent bronchioles and alveoli, a possibility of intralobar sequestration was considered. At this point the clinical and gross pathologic data were used

to confirm the possibility of a postinfectious cysts in some cases. The presence of an aberrant artery arising from the aorta and entering the area of the cyst was taken as definite evidence of intralobar sequestration, and these cases were excluded. Thus, the remaining intrapulmonary cysts showed little inflammation of the cyst wall and were surrounded by essentially normal parenchyma (Fig. 9A). The presence of squamous epithelium and associated smooth muscle cartilage plates and seromucinous glands was noted for each of the remaining cases.

In the mediastinum several classifications of congenital cysts have been proposed. There is general agreement that cysts that have gastric epithelium in whole

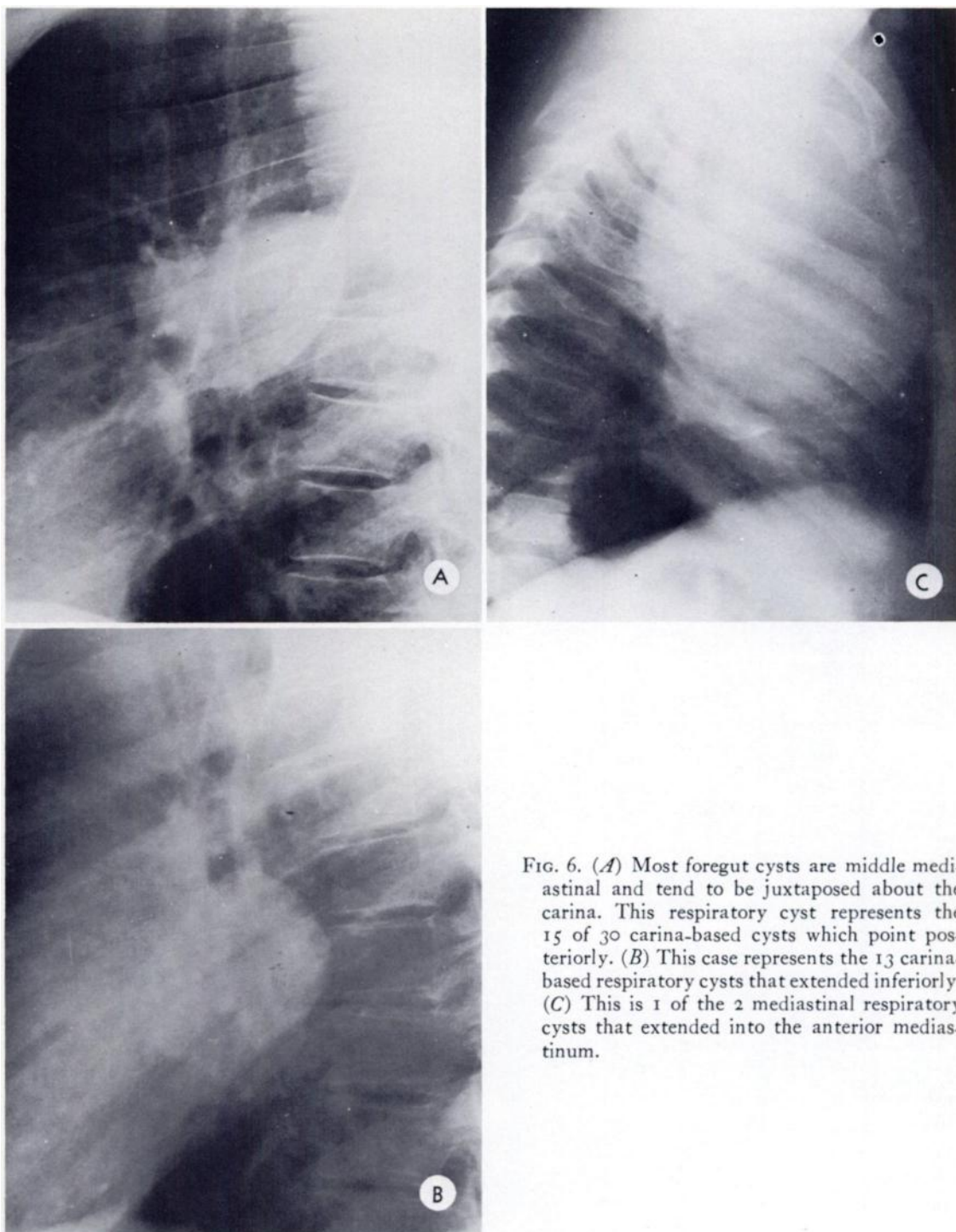


FIG. 6. (A) Most foregut cysts are middle mediastinal and tend to be juxtaposed about the carina. This respiratory cyst represents the 15 of 30 carina-based cysts which point posteriorly. (B) This case represents the 13 carina-based respiratory cysts that extended inferiorly. (C) This is 1 of the 2 mediastinal respiratory cysts that extended into the anterior mediastinum.

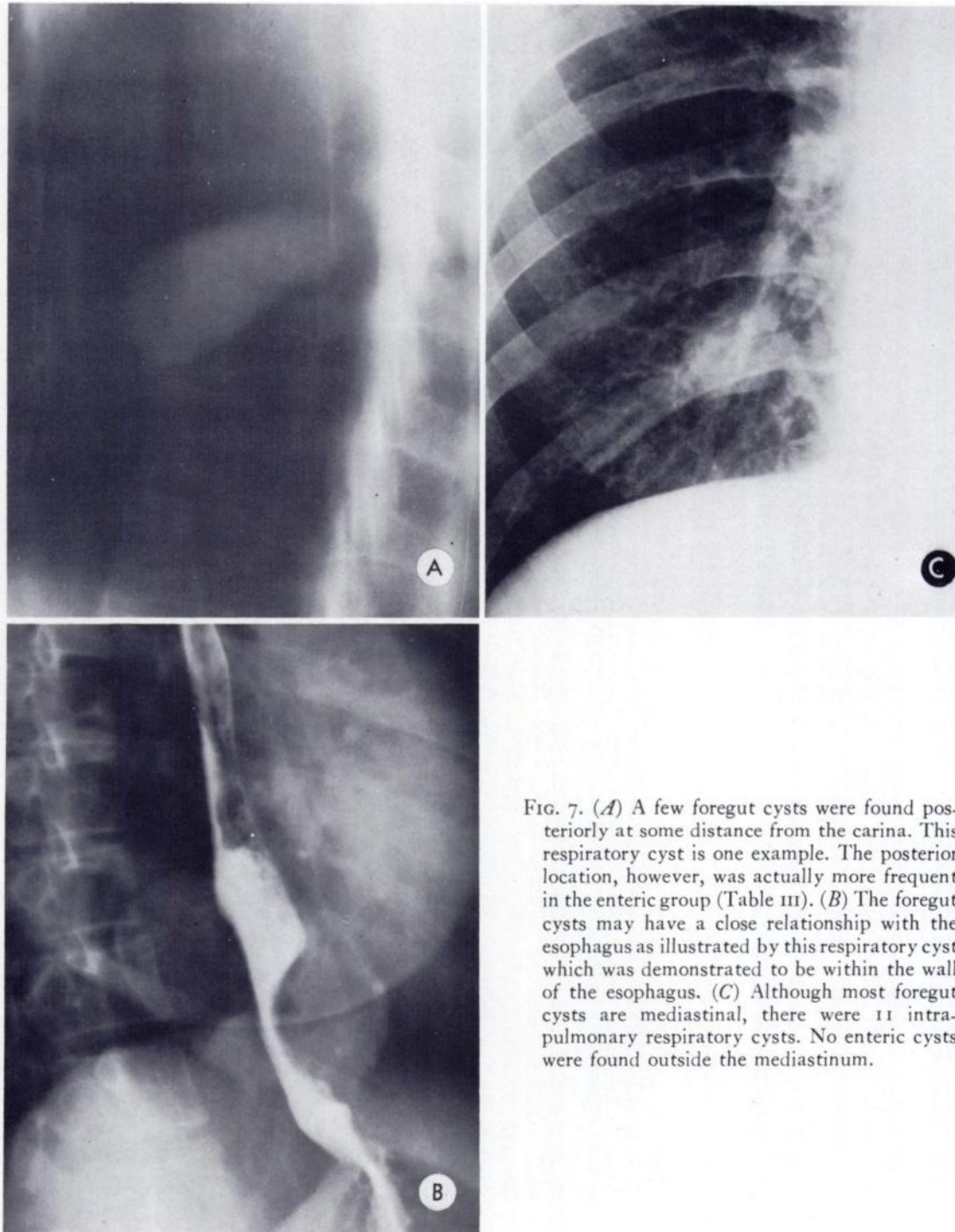


FIG. 7. (A) A few foregut cysts were found posteriorly at some distance from the carina. This respiratory cyst is one example. The posterior location, however, was actually more frequent in the enteric group (Table III). (B) The foregut cysts may have a close relationship with the esophagus as illustrated by this respiratory cyst which was demonstrated to be within the wall of the esophagus. (C) Although most foregut cysts are mediastinal, there were 11 intrapulmonary respiratory cysts. No enteric cysts were found outside the mediastinum.



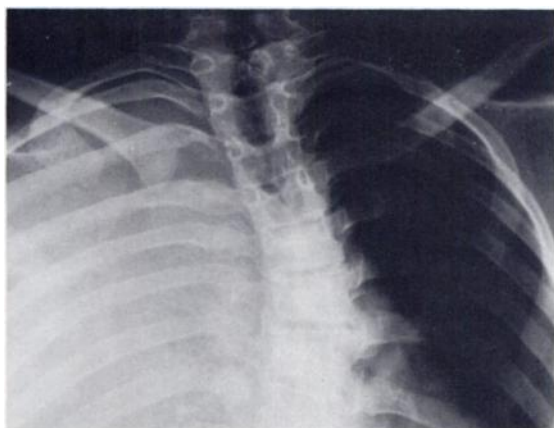


FIG. 8. Associated vertebral body anomalies were observed in 6 enteric cysts. Such cysts were then properly identified as *neurenteric*. This was the only case with a cervical spine anomaly and it should be emphasized that the spinal anomaly may be located *above* the level of the cyst itself.

or in part, represent a distinct type and should be classified as (gastro) enteric (Fig. 9B). Mediastinal cysts containing cartilage were considered definitely as respiratory (bronchial or bronchogenic) (Fig. 9C). Those with seromucinous glands were considered as probably, although not definitely, respiratory because a seromucinous type of gland occasionally occurs in the esophagus. The remaining cysts were lined by respiratory epithelium without other distinguishing features (Fig. 9D). In the last type of cyst, the possibility that the cyst was of embryonic esophageal or undifferentiated foregut origin could not be excluded. However, no cyst lined by squamous epithelium in a manner simulating adult esophagus was seen, and no cysts had other histologic features which would strongly suggest a cyst of purely esophageal type. Because of the lack of recognition of a mediastinal cyst of true adult esophageal type, it is probable that most of the cysts in which cartilage is presently not identified either would contain cartilage in additional sections or represent a less differentiated form of the typical respiratory cyst. For this reason the cysts with and without cartilage have been classified together and considered as respiratory.

A review of the pertinent literature on congenital intrapulmonary respiratory cysts points out the fact that there are no absolute criteria to separate an acquired from a congenital cyst pathologically, although the absence of associated parenchymal and bronchial inflammation and the lack of a clinical history of pulmonary inflammation are suggestive of a congenital cyst.

#### SUMMARY

Eighty-six cases of thoracic foregut cysts were histologically selected and classified as either respiratory (bronchogenic or bronchial) or enteric. Most are homogeneous water density masses which are located in the mediastinum and frequently are based around the carina. There is a tendency for the carina-based group to extend either posteriorly or inferiorly into the retrocardiac area. These masses tend to be right-sided and partially obscured by surrounding normal structures. The simple enteric cysts are not always distinguishable from the respiratory cysts, but they do tend to be larger and more posterior. In contrast with the simple enteric cysts, the *neurenteric* cysts should be very characteristic because of their associated vertebral body anomalies.

The intrapulmonary cysts may present as either a mass or cavity and such an appearance may be nonspecific. However, when the diagnosis is considered either preoperatively or on histologic grounds, signs of an underlying inflammatory process, such as surrounding infiltrate, bronchiectasis, pleural scarring or pleural effusion are good evidence for seriously questioning the diagnosis of a congenital cyst.

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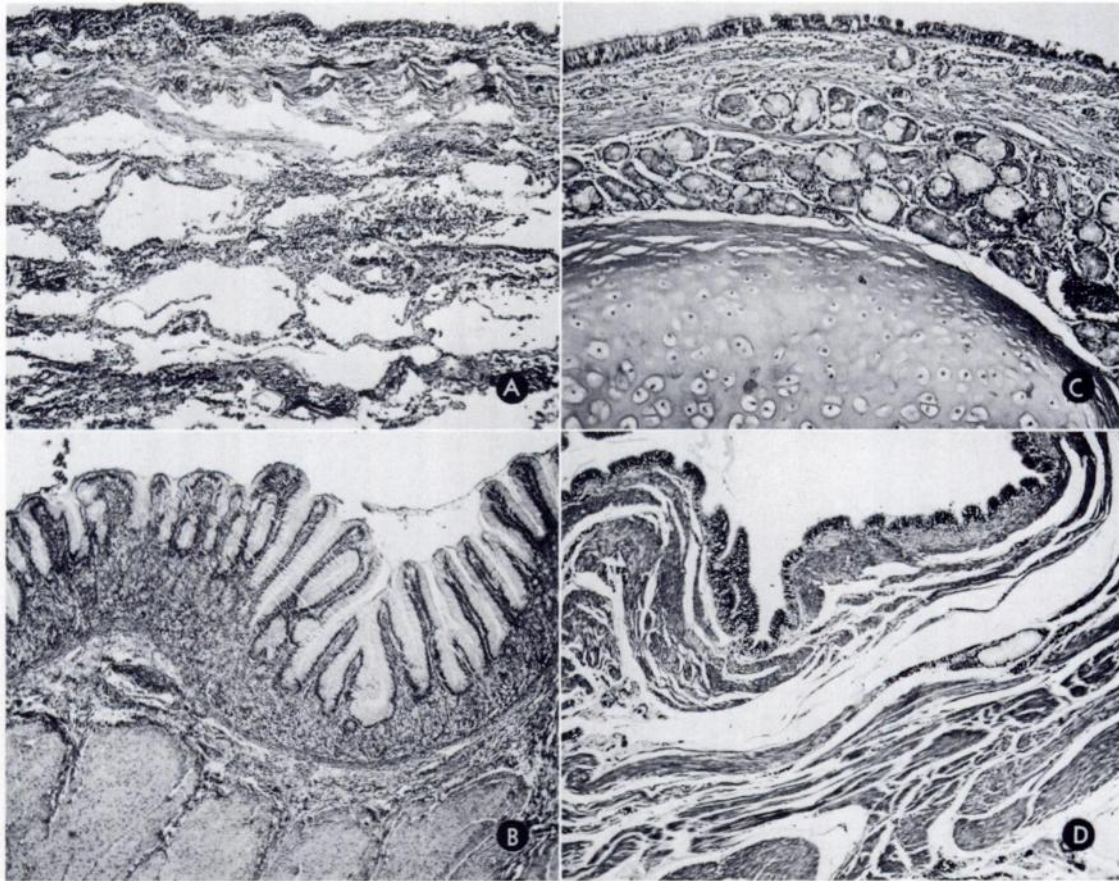


FIG. 9. (A) This histologic field from a peripheral bronchial cyst shows a thin fibrous wall lined by respiratory epithelium. Adjacent lung tissue is normal except for artifactual hemorrhage. (B) Mucus-secreting epithelium, gastric glands, and thick layers of smooth muscle are seen in this enteric cyst with gastric differentiation. Areas of esophageal differentiation are less common in these cysts. (C) This typical cyst of the mediastinum is lined by respiratory epithelium and contains cartilage and seromucinous glands in its wall. Squamous epithelium is occasionally seen lining such cysts. (D) This mediastinal cyst has respiratory epithelium and a wall containing only smooth muscle and rare mucous glands. Although included with the respiratory cysts, this one could be regarded as an embryonic esophageal or undifferentiated foregut cyst because of its intimate relationship with the esophagus.

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