The association of amyloid disease with multiple myeloma has been stressed by Magnus-Levy (1933). In the cases he described the amyloid either was localized or had affected the tissues characteristically involved in atypical amyloidosis. It is better, then, to speak of two types—primary or atypical, and secondary or typical. Indeed, the simple terms "primary" and "secondary" seem better. Primary amyloidosis occurs without any known cause, with the apparent exception of multiple myeloma, affects mesodermal tissue, and may be localized, forming a tumour, or generalized. It is possible that the deposit is not really true amyloid. Secondary amyloidosis follows certain well-known chronic diseases, and affects the abdominal organs, such as liver, spleen, and kidneys; true amyloid is present. Our case belongs to this secondary group, following as it does the chronic disease of rheumatoid arthritis.

The clinical findings in this case conform in general to those of the other reported cases. The history of enteritis at the age of 25 is interesting, as attacks of diarrhoea are common in these cases. The poor general condition with marked loss of weight is to be noted. The anaemia is also to be expected, but the tendency to haemorrhage is in some ways unusual. In most of the cases reported where haemorrhage has taken place the bleeding has been from organs affected by the amyloid process. Haematemesis, for instance, has occurred in systemic amyloidosis of the alimentary tract. In this case bleeding started sixteen months before death and took the form of a severe menorrhagia. When in hospital haematemesis occurred, ending in a terminal severe bleeding from the gums. None of the organs concerned had amyloid deposits. The liver and spleen were clinically enlarged. At necropsy the spleen had contracted and its weight was within normal limits. The liver, however, was definitely waxy and slightly enlarged. Albuminuria is generally regarded as a definite sign of amyloid disease of the kidneys but is not always present. In our case it appeared to be intermittent. This is very surprising in view of the state of the kidneys and the raised blood urea.

Summary

A case of amyloid disease complicating rheumatoid arthritis is reported in a female aged 28.

Several unusual clinical and pathological features of the case are noted.

The post-mortem findings are detailed, and a full report of the histological examination, with illustrations, is given.

The aetiology of amyloid disease is discussed, and a simple classification into "primary" and "secondary" is suggested.

We are indebted to Mr. T. C. Dodds, F.R.P.S., F.I.B.P., for the illustrations.

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A useful guide to current French medical literature is published by Editions Cartier, 13, rue Puits-Gaillot, Lyon, in the form of a journal entitled *Diagnostics et Traitements*. Each number contains brief abstracts in French of articles arranged in alphabetical order according to subject. For example, a current number contains abstracts on subjects from "F" to "H," beginning with "ferments" and ending with "hysteria." Presumably the entire alphabet is covered in a year. The majority of articles are from French sources, though foreign literature receives some attention. Some original and review articles are also scattered through the text. Two special journals are also published by Editions Cartier, *Revue de Diététique* (monthly) and *Archives de Rhumatologie* (quarterly). In these greater emphasis is laid on publication of original articles, although abstracts of current literature are included.

CONGENITAL DEFECT OF LEFT DIAPHRAGM WITH VOLVULUS OF STOMACH AND TRANSPOSITION OF VISCERA

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[WITH PHOTOGRAVURE PLATE]

Whilst the various abnormalities which may cause the abdominal organs to be placed in the thoracic cavity have been described by many observers, much confusion exists on the subject, the different types of anomaly being so mixed that a careful study of the reported cases often leaves one in doubt as to which one the author is describing. The clearest classification of the various diaphragmatic abnormalities is that given in several articles by LeWald (1924, 1927, 1928). There are true congenital diaphragmatic hernia in which developmental defect of varying size occurs and the hernia is contained within a sac; false diaphragmatic hernia, either congenital or traumatic, in which no sac is present; eventration of the diaphragm, in which an attenuated half of the diaphragm is arched so high that the abdominal organs are in close proximity to the heart; congenital absence of half of the diaphragm; and the rare condition of thoracic stomach, which, according to Bailey (1919), is a term used to indicate the presence of the stomach in the chest cavity without evidence of any abnormal opening in the diaphragm. The shortened oesophagus opens into a stomach placed above the diaphragm, and the pylorus or duodenum passes through the oesophageal opening in the diaphragm.

The case described below is of particular interest by reason of (1) the difficulty in establishing by radiology the exact size and nature of the congenital defect, and whether the condition was amenable to surgical intervention; and (2) a sudden and inexplicable volvulus of stomach together with equally sudden transposition of viscera—resulting in death, which occurred while the case was under review—whereby the herniated small and large intestines underwent spontaneous reduction into the abdominal cavity, and the dilated stomach, which had previously been normally placed in the abdominal cavity, became transposed into the thorax.

Case Report

The patient, a male infant aged 8 weeks, was admitted to the Cardiganshire General Hospital, Aberystwyth, on Sept. 4, 1946, stated to be suffering from congenital pyloric stenosis. There was a history of normal delivery, the weight at birth being 6½ lb. (3.06 kg.). This was the mother's second pregnancy, and a sister aged 2 years was healthy and free from known congenital defects. The mother had noticed that the breathing was very difficult from birth. There were frequent attacks of cyanosis with aggravation of dyspnoea, which appeared to be worse after feeding. The baby was bottle-fed, and vomiting was an early symptom. Sometimes the vomit occurred immediately after the feed; at other times it was delayed and projectile. Invariably the act of vomiting relieved the cyanosis and dyspnoea. The mother had found that nursing usually aggravated the dyspnoea, and that the most comfortable position appeared to be lying down. On admission the baby's weight was 7 lb. 12 oz. (3.5 kg.); its colour was normal; the respiration

rate was 72, pulse rate 150-160, and temperature 101° F. (38.3° C.). There was a suggestion of asymmetry of the chest wall, with the left side possibly a little more prominent than the right. There was a dull tympanitic note to percussion over the whole of the left chest back and front, with absent breath sounds and occasional harsh grating sounds, together with numerous gurgling sounds somewhat similar to but not identical with those of intestinal peristalsis. The maximum intensity of the heart sounds was in the nipple line of the right thorax. The day after admission he was referred for radiography of chest.

An A.P. film of the chest showed a marked displacement of the heart to the right side. The whole of the left side of the thorax showed loculated radiolucent shadowing and there was a complete absence of any normal lung pattern. The medial half of the left dome of the diaphragm appeared normal in position, but the outline of the lateral half could not be defined. The air bubble in the cardiac end of the stomach was seen below the diaphragm and showed a normal rounded outline. The appearances were suggestive of a cystic lung condition or the presence of bowel in the thorax. Later the baby was given a barium meal of about 3 oz. (85 g.). The oesophagus was displaced to the right, but appeared to open into the cardiac end of the stomach in the normal position. The stomach was large but showed good contraction, and was empty in about four hours. The duodenum led direct into the thoracic cavity at a point just lateral to the centre of the left dome, and it appeared to be taut. The left side of the thorax was later seen to contain all the remaining bowel except part of the descending colon leading down to a normally situated rectum (Plate, Fig. 1). The whole of the meal was in the large bowel at 24 hours and almost completely evacuated at 48 hours. On screening, the right dome of the diaphragm moved normally. The diaphragmatic movement on the left side, as outlined by the barium-filled bowel and upper margin of liver, was limited but was not paradoxical.

The patient was given small feeds of humanized milk, and with careful nursing but without the adoption of any special measures his condition gradually improved, and within four weeks of admission he was taking 5-oz. (140-g.) feeds three-hourly. There was occasional slight vomiting and the post-prandial dyspnoea remained a marked feature, but he continued to thrive. An attempt was made to keep him at an angle of 45 degrees, but it was noticeable that this aggravated his distress and that the most comfortable position was the recumbent one.

On Feb. 23, 1947, at the age of 7 months, he was taking 8-oz. (225-g.) feeds three-hourly, alternating with groats, custards, etc., and his weight was 14 lb. (6.35 kg.). He was an extremely contented infant provided he had his feeds regularly, and he raised no objection to repeated x-ray examinations. A detailed examination of his condition on this date revealed the following: The cardiac rate was about 150 a minute, with maximum intensity in the right nipple line; the respiration rate 62. There was marked asymmetry of the chest, the left side being more prominent. The circumference of the right half of the thorax was 8½ in. (20.6 cm.) and of the left half 8½ in. (21.9 cm.). There was an exaggerated inspiratory divergence of the left costal margin from the median line in the epigastrium, with consequent greater widening of the left than the right side of the subcostal angle, and there was inspiratory retraction of the epigastrium. Breath sounds were absent over the whole of the left chest, and there was an abundance of gurgling sounds. Breath sounds appeared to be normal on the right side. The measurement of the abdomen was 14½ in. (36.18 cm.), and its contour appeared normal; the liver dullness extended downwards for two fingerbreadths below the right costal margin. For the week commencing Feb. 23 he had a bout of diarrhoea. He refused to take his full feeds, and vomiting, non-projectile in nature, occurred more frequently. On Feb. 26 he was given another barium meal. This showed a considerable increase in the size of the stomach but otherwise no change. The position of the small and large intestine had remained constant over a period of five months (Plate, Fig. 2). He continued to be reluctant to take his full feeds, there was occasional vomiting, and he lost a little weight. On March 7, in conjunction with the other infants in the ward, he developed rubella, and was transferred to the Isolation Hospital. We were informed that his condition there did not give rise to anxiety until March 11, when

he was very fretful and reluctant to take his feeds. After a 4-oz. (114-g.) feed at 10 p.m. he appeared to be in definite pain. Nursing and rubbing his back seemed to comfort him, but with an increasing respiration rate he died at 3 a.m. on March 12.

Post-mortem Findings

We are indebted to Dr. M. V. N. Sudds for the necropsy report:

"The whole of the small and the large bowel was situated in the abdominal cavity together with the spleen and the liver, which was greatly enlarged. The right kidney was congested. The duodenum was in its normal situation. An enormously distended stomach occupied the whole of the left and encroached on the right thorax. There was a volvulus of the stomach, the pyloric end having rotated a half circle, so that the stomach was inverted, with its posterior surface facing forwards and its greater curvature lying underneath the first rib, and with its fundus resting on the thoracic aspect of what was present of the left diaphragm (Plate, Fig. 3). The oesophageal opening in the diaphragm was normally placed, but the abdominal portion of the oesophagus was elongated, and it curved to the left and upwards around the right margin of an abnormal opening in the posterior half of the left diaphragm, so that the cardiac orifice lay within the thoracic cavity.

"Adjoining the abdominal portion of the oesophagus as it entered the hiatus was the twisted and elongated pyloric end of the stomach (Plate, Fig. 4). In the illustration the liver and spleen have been removed and the intestines drawn aside. The stomach had to be aspirated of a large portion of its contents before it could be reduced into the abdominal cavity. It contained 450 ml. (16 oz.) of milky fluid in addition to an abundance of gas. Partly emptied of its contents, it measured 5½ in. (13.6 cm.) from fundus to greater curvature, and its greatest width was 4½ in. (11.1 cm.).

"The heart lay well over in the right thorax, with a small strip representing the left lung compressed between the mediastinum and the right edge of the stomach, and with the right lung situated in, but by no means filling the remainder of, the right thorax (Plate, Fig. 3). In view of the fact that the weight of the left lung was only 12 g. (2/5 oz.) and that it was completely airless and sank in water, the size of the right lung was surprisingly small. From apex to base it measured 4½ in. (11.5 cm.), its base was 3½ in. (9.94 cm.), its maximum thickness 1½ in. (3.17 cm.), and its total weight 87 g. (3 oz.). There was a large oval-shaped opening in the left diaphragm measuring 2½ in. by 1½ in. (5.7 by 2.85 cm.); it admitted six fingers, and occupied most of its posterior half. Its right edge was situated about 3/4 in. (1.9 cm.) from the oesophageal opening, and its right margin curved to its origin from the medial lumbo-costal arch. Its left margin was curved and ended as a thin strip attached to the inner surfaces of the ribs. The anterior half of the left diaphragm appeared to be strong and well developed. The edge of the hiatus was lined with serous membrane so that the peritoneum and parietal pleura formed a continuous sheet."

Embryology

The hernial opening in this case was the result of lack of closure of the pleuro-peritoneal hiatus of the diaphragm, and a better understanding of the nature of the defect is obtained from the following description of the development of the diaphragm by Shanks (1938).

"In the early embryo the pleural cavities, protrusions of the primitive coelom, communicate freely with the peritoneal cavity. The diaphragm which eventually separates them is a composite structure embryologically (see Diagram). The ventral half is formed from the septum transversum, a mesodermic partition separating the heart from the abdominal

Diagram showing components of the diaphragm: 1, Aorta; 2, Oesophagus; 3, Medial mesentery; 4, Inferior vena cava; 5, Pleuro-peritoneal folds; 6, Septum transversum.

viscera. It first lies obliquely in the neck, but gradually moves down and assumes a transverse disposition. It reaches its final position opposite the level of the 12th rib at about the fourth week. In its descent downwards it carries with it its nerve supply from C.3, 4, and 5—the phrenic nerve. The point of entry of the latter into the adult diaphragm marks the posterior limit of the portion formed from this septum. The posterior half of the diaphragm is formed from three structures: the primitive mesentery of the foregut, stretching between the dorsal coelomic wall behind and the septum transversum in front; and the pleuro-peritoneal folds, one growing inwards from each lateral coelomic wall to fuse with the septum transversum and the medial mesentery. The last portion to fuse is the postero-lateral, where for a time there persists, on each side, the pleuro-peritoneal canal or hiatus. Failure of this hiatus to close accounts for a common type of hernia."

Discussion

Subcostal Angle.—According to Lord (1927), the observations of Hoover (1913, 1919, 1920a) on the functions of the diaphragm suggest that in the presence of eventration important evidence may be obtained by comparison of the respiratory movement of the two sides of the chest. His clinical and experimental observations indicate that the diaphragm acts as an antagonist to the intercostal muscles, which enlarge the transverse and antero-posterior diameter of the base of the thorax and widen the subcostal angle. According to him, the extent to which the diaphragm is capable of opposing the intercostal muscles depends on its arch, and the more nearly the curve approaches a plane the greater its mechanical advantage. If the diaphragm is at a mechanical disadvantage by upward displacement or if its musculature is impaired, the pull of the intercostal muscles is accentuated. Lord (1927) reports two cases of eventration of diaphragm which presented exaggerated inspiratory widening of the subcostal angle due to a greater divergence of the left costal margin, and he quotes Funk and Manges (1920), Korns (1921), and Reifstein (1925) as having made similar findings. Hoover (1920b) attaches equal importance to this physical sign, and he states that the observation of increased movement of the costal margin on the affected side is of service in the differentiation between hernia and eventration, and that in hernia the movement of the costal margin is not modified. Korns (1921) explains the absence of this physical sign in a case of hernia on the grounds that in this condition the diaphragm is not aplastic and in a high position, but has intact muscles and an approximately normal position. Their writings suggest that these authors regard it as being diagnostic of eventration of diaphragm. The clinical findings in this case appear to be at variance with their views, since the exaggerated inspiratory divergence of the left costal margin was a marked feature in the case we quote, as also were the inspiratory retraction of the epigastrium and increase in circumference of the chest on the affected side. It would thus be reasonable to conclude that the extent to which the diaphragm is capable of opposing the intercostal muscles does not depend solely on the level and strength of its arch, but that the accentuated pull of the intercostal muscles will produce these physical signs if the diaphragm is at a mechanical disadvantage by reason of impaired musculature from any cause.

Mode of Death.—It can be inferred that the cause of death was cardiac embarrassment caused by the pressure of the twisted, obstructed, and distended stomach. What was the sequence of events which led up to the fatal condition? During his five months' stay in hospital a series of five x-ray examinations were carried out, and in all of them the relative position of the stomach and bowel had remained constant. What was the factor which suddenly induced such a complete transposition of these organs?

(a) Did the herniated intestines first undergo spontaneous reduction into the abdominal cavity and then the increased intra-abdominal pressure result in the stomach being forced into the thorax and in so doing undergo a volvulus? This would not appear to be likely, since in none of the films taken during this period was there any evidence of the slightest change in position of the small and large intestines within the thorax, or of the descent of any portion of them into the abdominal cavity. It would be difficult to explain why they should suddenly decide to migrate spontaneously on this particular day.

(b) A more reasonable explanation would be that the partially emptied stomach underwent a volvulus and insinuated itself through the abnormal opening into the thorax, and that for a short time it remained in the thorax with the intestines. Then when the stomach, with its pyloric end obstructed, became more distended as the result of further feeding and fermentation, its presence in the thorax gradually forced down the intestines into the abdominal cavity.

(c) If the latter was the case, was the increase in size of the stomach that was noticeable in the last series of films of any significance? And did the apparently taut first part of small bowel running obliquely upwards into the thorax as shown in Fig. 2 play any part in the causation of the volvulus?

Summary

A case of hernia through the pleuro-peritoneal hiatus is described.

It is shown that the clinical signs of exaggerated inspiratory excursion of the costal margin with consequent greater widening of the subcostal angle, together with inspiratory retraction of the epigastrium on the affected side, are not to be regarded as diagnostic of eventration of the diaphragm, but as evidence of imperfect diaphragmatic function by reason of impaired musculature from any cause.

An untimely death, with subsequent necropsy, revealed an unexpected interchange of position of the stomach and intestines, with a volvulus of the former as the cause of death.

An attempt is made to outline the sequence of events which led up to this volvulus and transposition of viscera.

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Speaking on the early diagnosis of pulmonary tuberculosis at a clinical meeting of the East Sussex Medico-Chirurgical Society held at St. Leonards-on-Sea on June 24, Dr. E. A. Wood said that he wanted to make a few suggestions on discovering cases while they were still curable—preferably before either symptoms or cavitation had developed. His first suggestion was to encourage routine x-ray examination of the apparently healthy. So also they should arrange for the x-ray examination (and, in young children, tuberculin testing) of every household contact of a case of pulmonary tuberculosis. Furthermore, disease would not necessarily show itself on first examination; contacts must be x-rayed—not less often than annually—over a period of years, particularly at those times of life, such as the late teens and early twenties, when the disease seemed most likely to appear. Saying that in February they went into the surgery and found twenty or thirty patients with coughs, he suggested the following cases most needed x-ray examination of the chest: (1) All cases of haemoptysis; (2) all cases of pleurisy (dry or wet) not obviously due to acute pneumonia; (3) patients convalescent from any serious "chest" illness; (4) those who had had any chest illness not clearing up in the expected time; (5) anyone still with a cough after efficient treatment for four weeks; (6) cases of vague ill-health not yielding to ordinary treatment; (7) cases of dyspepsia not yielding to ordinary treatment; (8) patients who lost weight without other cause being found. As soon as his trouble had been diagnosed, the tuberculous patient should be put strictly to bed while he awaited sanatorium treatment. "Avoid that futile, ambiguous, but oft-repeated instruction, 'Go home and take things quietly, and come and see me in two months' time.'"

CONGENITAL DEFECT OF LEFT DIAPHRAGM WITH VOLVULUS OF STOMACH AND TRANSPOSITION OF VISCERA: E. OWEN-LLOYD AND DEINIOL ROBERTS

FIG. 1.—Lateral radiograph after barium meal, showing large intestine in thorax. (Sept. 17, 1946.)

FIG. 2.—Small intestine leading into thorax appears taut. Considerable increase in size of stomach. (Feb. 26, 1947.)

FIG. 3.—Note size of liver and position of greater curvature.

FIG. 4.—Opening in diaphragm from below.

ACUTE INTERSTITIAL POLYMYOSITIS TREATED WITH PENICILLIN: I. FRIEDMANN AND F. POR

FIG. 1.—Section of sternomastoid before penicillin therapy, showing extensive inflammatory changes ($\times 160$).

FIG. 2.—Same muscle fourteen days after treatment. Note disappearance of inflammation.