were consistently acted upon the mortality of diphtheria would be considerably higher than it is at present.

In a series of 3,000 diphtherial patients of whom I was in charge throughout their illnesses no fewer than 539, or 17.9 per cent., were admitted after the fifth day. With the exception of 12 very mild cases all the patients in whom membrane was present in the throat or nose received antitoxin. Of these 19 died—a mortality of only 5.5 per cent. As the great majority of these cases were in children, and every form of the disease was represented, the low mortality cannot be attributed to the patients being adults, among whom the death rate from diphtheria is usually low, nor to the mildness of the attack. In view of the fact that in the pre-antitoxin era, even when cases were brought under treatment at the very onset of the disease, the mortality was never less than 28.8 per cent., and often rose to 50 per cent. or higher, we can only ascribe the low figure of 5.5 per cent. to the use of antitoxin.

It is important to realize, as I pointed out many years ago (1907), that the evolution of diphtheria may be rapid and malignant, so that even enormous doses of antitoxin given on the second day of disease may not avert a fatal issue; or comparatively slow and benign, so that the disease, which may have been in progress for a week, yields rapidly to a small dose of antitoxin without any sequels.

We should, therefore, be guided by clinical rather than chronological considerations in our treatment of diphtheria—the presence of membrane in the throat, nose, or elsewhere, however late in the disease, being an indication for serotherapy. I have seen so many late cases in which my original gloomy prognosis was happily falsified that I am inclined to say of diphtheria as Ricord did of syphilis: "As regards recovery, everything is possible, sometimes even the impossible."

The chief advance in recent times in the treatment of diphtheria is the use of a refined or concentrated serum whereby the incidence of rashes and other unpleasant sequels is reduced to a minimum. Since our employment of this serum at the Western Hospital in April, 1927, the frequency of serum sickness has fallen from about 60 to under 30 per cent. The only serum phenomenon now seen is urticaria, which is often limited to the site of injection, lasts only for a few hours, and is not accompanied by rise of temperature or constitutional disturbance. With the raw or unconcentrated serum not only was urticaria much more frequent, but in about 12 to 15 per cent. of the cases injected a secondary rash, at first amorphous or morbilliform and later circinate, appeared, usually attended by a more or less considerable rise of temperature, pains in the joints or muscles, and a varying degree of cervical adenitis. In a small percentage of cases there was a return of the sore throat, manifested by patches on the tonsils, to which I gave the name many years ago (1906) of "angina redux." Since the refined serum has been employed I have not seen any of these manifestations of the secondary syndrome.

The introduction of this refined serum is a great boon to the patient. Though somewhat more expensive than the unconcentrated antitoxin it is well worth the additional cost. In previous times it not infrequently happened that the patients, especially adults, suffered as much or even more from serum sickness than from an attack of diphtheria.

General Treatment.

Next to early administration of antitoxin the most important principle in the treatment of diphtheria is the enforcement of rest for a period proportionate to the severity of the initial angina. In mild cases rest in bed for the greater part of the time in the recumbent position should continue for three weeks, while in severe cases the period of recumbency should be extended to six weeks, or more if any paralysis is present.

It is doubtful if any drug has the power to modify favourably the course of the disease, though I am inclined to think that adrenaline may be of value in forestalling or combating the suprarenal insufficiency which plays an important part in the syndrome known as cardiac paralysis.

Alcohol was long regarded as an indispensable drug in the treatment of diphtheria, and as such was advocated by such well-known authorities as Jacobi of New York

and Ker of Edinburgh, as well as by many lesser luminaries in the medical firmament. Even now it is still used on a large scale in the acute stage or in convalescence by some pyretologists, who harbour the delusion that alcohol is a cardiac stimulant or is otherwise indispensable.

When I was appointed medical superintendent at the Western Hospital I found that the annual consumption of brandy in the previous year (1925) had been 2,589 oz., most of which had been employed in the treatment of diphtheria. I at once set to work to convince my colleagues that alcohol, so far from being of any value in the disease, was liable to increase the myocardial degeneration initiated by the diphtheritic poison.

The result of my anti-alcoholic campaign was that the amount of brandy ordered on the diet sheets fell from an average of about 30 oz. daily to *nil* without any ill effects, the diphtheria mortality being lower in 1926 (4.53 per cent.) than in the previous year (8.54 per cent.), while in 1927, when no alcohol at all was used in the treatment of the disease, the diphtheria mortality (3.01 per cent.) was lower than in any of the other Metropolitan Asylums Board hospitals, although the type of disease has remained the same during the last three years.

Time does not permit me to deal with the relative merits of tracheotomy and intubation, or to discuss the management of diphtheritic paralysis, about which there is little new to be said. A few remarks, however, may be made, in conclusion, as regards the local treatment of diphtheria. Since the introduction of antitoxin local treatment in the form of syringing, spraying, or gargling has occupied a very secondary position in treatment. In my series of 3,000 cases I have employed local treatment in only 364 cases, or 12.13 per cent. Most of these patients were older children or adults, who welcome any form of local treatment, which young children as a rule resist.

From comparative observations I have found that the average period for the throat to become clean after injection of antitoxin is practically the same, whether local treatment is employed or not.

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RARE COMPLICATIONS OF ACUTE APPENDICITIS.*

BY

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THE following cases of appendicitis are of considerable interest and seem to deserve recording. They are noteworthy on account of their extreme rarity, and because they present many unusual features, which increased the difficulty of pre-operative diagnosis.

CASE I.—Acute Appendicitis Accompanying a Perforated Duodenal Ulcer.

A man, aged 35, was admitted to hospital on August 21st, 1927, complaining of abdominal pain and vomiting.

History.—Nine hours before admission the patient was seized with sudden pain in the upper part of the abdomen. The pain started slightly to the right of the mid-line, and passed down the right side of the abdomen into the right iliac fossa. It was sufficiently severe to double him up, and its onset was accompanied by vomiting. The patient had suffered off and on for eighteen years with pain in the upper abdomen, coming on two hours after food and lasting for a variable period. The pain was relieved by taking food. He gave no history of vomiting, and had never been jaundiced; the bowels and micturition had always been normal.

* From a paper read to the Kidderminster Medical Society, April 20th, 1928.

Examination.—On admission the patient looked ill; the mucous membranes were slightly cyanosed, the tongue was dry and furred. Temperature 98° F., pulse 110, respirations 24. The pupils were semi-dilated, and reacted normally to light and accommodation. Nothing abnormal was found in the examination of the chest. The abdomen did not move on respiration, and there appeared to be some fullness in the right iliac fossa. There was tenderness and rigidity all over the abdomen, which was most marked in the right iliac fossa. There was dullness in the flanks and in the right iliac fossa. Liver dullness was present. Rectal examination disclosed nothing abnormal.

Operation.—An immediate exploratory laparotomy was performed, a two-inch incision being made in the right iliac fossa. On opening the peritoneal cavity much turbid but odourless fluid exuded. The appendix was brought into the wound, and was found to be swollen; the whole of the outer coat was markedly inflamed, and there were many recent adhesions around it. The appendix was removed in the usual manner. On further examination it was discovered that the lumen was filled with a thick blood-stained fluid, which had a fetid odour. On washing away the fluid it was seen that the mucous membrane of the appendix was altered in its distal half, and that this region was studded with multiple large thrombi. No perforation was present. It was not thought that the condition of the appendix was sufficient to account for the large amount of free turbid fluid in the peritoneal cavity. A second right paramedian incision was made in the upper abdomen, when it was discovered that turbid fluid was oozing from a small punched-out perforation, one-eighth of an inch in diameter, situated on the superior surface of the duodenum, just distal to the pylorus. An indurated area about one inch in diameter was palpated, surrounding the perforation. The perforation was closed by a purse-string suture and then sewn over. The gall-bladder was found to contain a hard mass, but no operation on it was performed. The upper incision was partially closed with tubal drainage down to the region of the perforation. A drainage tube was passed through the incision in the right iliac fossa down into the pelvis, and the rest of the wound was closed.

The patient made an uninterrupted recovery and was discharged from hospital three weeks later with his wounds healed; he was advised to return to hospital in one month for exploration of the gall-bladder.

He was readmitted one month later, and cholecystostomy was performed, when the gall-bladder was found to contain a mass of clay-like material; but there were no definitely formed stones. The cystic duct was full of the same material, which was evacuated with a gall-bladder spoon. A tube was inserted into the gall-bladder and secured with a purse-string suture. The gall-bladder was then sewn to the posterior aspect of the anterior abdominal wall and the rest of the wound was closed. There was slight bile drainage for four weeks, after which the patient was discharged from hospital quite well, with his wound healed.

Comment.

The association between duodenal ulcer and chronic appendicitis has long been well recognized, but perforation in the former and acute inflammation in the latter occurring at the same time is very unusual, although it is not infrequently found that some degree of peri-appendicitis accompanies a perforated ulcer.

Where two conditions are present at the same time it is always wise to associate them rather than to make two separate diagnoses; nevertheless, in this case the possibility must be borne in mind that the duodenal ulcer had been present for a number of years, and had no intimate connexion with the acute inflammation in the appendix. On the other hand, it may well be that there had been chronic inflammation in the appendix, which had been superseded by acute inflammation, and that this had caused the onset of an acute process in the ulcer previously present, which had resulted in perforation, or that the acute process in the appendix was secondary to an acute process in the ulcer.

In this case, although the symptoms pointed to its being one of duodenal perforation rather than acute appendicitis, the onset of pain being in the upper abdomen and there being a long history of indigestion, nevertheless, from the examination of the abdomen, some difficulty in pre-operative diagnosis was presented. The case was further complicated by the additional presence of gall-stone sand.

CASE II.—An Acute Gangrenous Perforated Appendix in the Sac of a Strangulated Inguinal Hernia.

A man, aged 77, was admitted to hospital on January 23rd, 1928, complaining of abdominal pain and vomiting.

History.—The patient had had bilateral inguinal hernia for forty years. Both herniae had always been easily reducible, and had been partially controlled by a truss for many years. Two days before admission he was seized with sudden severe pain in the right groin; the onset of the pain was accompanied by vomiting. The pain and vomiting had persisted, and the hernia on the right side had become tender and irreducible. The vomit had been dark brown in colour during the last twenty-four hours before admission. In the course of the previous two days his abdomen had gradually increased in size, and he had had

diarrhoea. For the last twenty-four hours he had not passed any flatus. Micturition had always been normal.

Examination.—On admission the patient looked ill. His tongue was dry, and covered with a dark brown fur. Temperature 99.6° F., pulse 110, respirations 24. Nothing abnormal was discovered in the examination of the chest. The abdomen was markedly distended, but moved on respiration. There was tenderness all over the abdomen, but no rigidity; the percussion note was resonant, and there was no dullness in the flanks. Bilateral inguinal herniae were present, extending down into the scrotum. The skin over the right hernia was reddened in the region of the external abdominal ring. The hernia was very tender on palpation, and the percussion note over it was resonant. No attempt was made to reduce the hernia. By the rectum nothing abnormal was discovered.

Operation.—An incision was made over the right hernia, the sac was defined, and the cord was separated. A small amount of green turbid fluid escaped on opening the sac, which was found to contain the caecum and omentum. The caecum was injected, and had recently formed flakes of lymph on its surface. The appendix was in the centre of the sac; the whole of its wall in its distal three-quarters was gangrenous, and there was a small perforation about one inch from the distal end. The gut was constricted by the neck of the sac. The cavity was swabbed out and the appendix was removed. The caecum and omentum were returned into the peritoneal cavity. The sac was tied and excised, and the wound closed with drainage.

Subsequent History.—The patient was treated with enemata and aperients. On January 24th he had a good result from a turpentine enema, passing much flatus and formed faeces. His abdomen was less distended and he had not vomited. The following day he had not passed flatus and the abdomen was more distended; he vomited green fluid several times during the day, and had clear results from enemata. On January 27th the abdomen was still more distended; he had not passed flatus, and had had clear results from enemata. He was vomiting dark brown fluid. The abdomen was not rigid.

Second Operation.—Exploratory laparotomy was performed through an incision in the right iliac fossa. On opening the peritoneal cavity it was found that the small intestine and caecum were markedly distended. There was no peritonitis. A band was felt constricting the large intestine just distal to the caecum, and the transverse colon and descending colon were collapsed. The constricting band was divided and caecostomy was performed. The wound was closed and the caecostomy was opened, when a slight amount of flatus escaped.

On January 28th the patient had frequent large openings of the bowels by the rectum. The caecostomy had not acted. Two days later he was much weaker. His bowels were well open both by the rectum and by the caecostomy. His abdomen was less distended. He was incontinent of faeces. He died on February 1st.

Comment.

In this case the question arises whether the condition was primarily one of acute appendicitis or whether it was secondarily due to strangulation of the appendix. The fact that there were recently formed flakes of lymph in the region of the appendix, and that the organ was perforated, suggests that the condition was primarily one of acute appendicitis rather than a strangulation of the appendix. An unusual feature in this case was the additional presence of a constricting band, which added considerably to the difficulties in pre-operative diagnosis, although, owing to the absence of signs of peritonitis, organic obstruction by a band had been suspected.

CASE III.—An Acute Gangrenous Perforated Appendix in the Sac of a Post-operative Ventral Hernia.

A man, aged 25, was admitted to hospital on February 1st, 1928, complaining of abdominal pain.

History.—Fourteen days before admission the patient had sudden severe pain in the right iliac fossa, accompanied by vomiting. The pain had persisted, although after the first few hours it had become much less severe, and was now merely a dull ache. He had had no further vomiting. He suffered from constipation, the bowels being opened with difficulty every two or three days. Micturition was normal. He had been operated upon two years previously for appendix abscess. The abscess was drained, but the appendix was not removed; he was advised to return to hospital three months later for removal of the appendix, but he failed to do so.

Examination.—On admission the patient looked ill; his tongue was moist and slightly furred. Temperature 98.4° F., pulse 82, respirations 20. Nothing abnormal was discovered in the examination of the chest. There was a ventral hernia about four inches in diameter in the region of the old appendix abscess scar; the skin over the hernia was normal. The abdomen moved well on respiration, and there was no distension, tenderness, or rigidity in any part. A hard mass 3 inches by 2 inches was felt in the centre of the ventral hernia; the mass was not tender, and there was no gurgling on palpation. The percussion note over it was dull. Nothing abnormal was discovered on rectal examination.

Operation.—An incision was made over the ventral hernia, and the sac was defined; it was thin-walled and lobulated, and contained a mass of omentum, in the centre of which was a gangrenous perforated appendix, surrounded by four ounces of pus. The appendix, surrounded by the omentum and pus, was wrapped in a gauze swab and removed. The scar tissue of the old wound was excised, and the fasciae and muscle were drawn

together with mattress sutures, the wound being partially closed with tubal drainage.

The patient made an uninterrupted recovery, and was discharged from hospital five weeks later, quite well, and with his wound healed.

Comment.

This was an undoubted case of a recurrent attack of acute appendicitis, and merely gives additional evidence to prove the inadvisability of not operating on these cases.

It is interesting to note that in this particular case the patient himself alone was to blame.

SYNDACTYLISM IN FOUR GENERATIONS.

BY

DAVID PERKOFF, M.R.C.S., L.R.C.P.

RECENTLY I have treated the rare condition of syndactylism in three generations of one family, and a member of the fourth generation, now deceased, was definitely known to have this deformity. These cases are submitted as an interesting problem on the subject of heredity.

Syndactylism, as the name implies, is a condition in which there are webbed fingers, with a more or less close union of contiguous digits. Three main varieties may be distinguished: (1) union by skin only; (2) union by skin and fibrous tissue; (3) a condition when the bones are more or less fused together.¹

FIG. 1

Photographs of the hands of the grandmother mentioned in the text. Fig. 1 shows the backs of the hands, and Figs. 2 and 3 the palms; the rudimentary separation of the thumb of the right hand is seen in Fig. 2.

FIG. 2.

FIG. 3.

Another variety, the "lattice type," exists in which the bases of the fingers are separated, but distally they are united. Such cases afford a strong reason for assuming a mechanical origin. It is possible that the fingers have developed, and then later, for some reason, they have been subjected to pressure and adhesion has taken place. This assumption is strengthened when it is noticed that sometimes at the point of union the last phalanx of one finger overlies the other. It is usually the inner side of the hand which is affected, and the fusion of the thumb and index finger is a rarity. Messrs. Roucayrol and Klausner have, however, reported cases of this rare type.

CAUSATION.

The leading theories of causation, according to Roucayrol and other authorities, are:

1. Lesions of the foetal nervous system (Guérin).
2. External pressure and traumatism during pregnancy (Cruveilhier).
3. Embryonic amniotic adhesion (Lommelougue).
4. Amniotic loops or bridles (Doreste).
5. Reversion to early conditions (Darwin).
6. Osler thinks it occurs more frequently in children of syphilitic appearance.

7. Edwards states that it is a stigma of degeneration occurring in persons of weakened intellect.

8. Goldman asserts that syndactylism may be regarded as an arrest of development, since during foetal life the fingers are bound together for a time by webs of varying extent, the thumb almost always remains free, and in most instances two fingers only, usually the third and fourth, are bound together.

Like many foot and hand conditions, syndactyly is markedly hereditary.² Luaces³ mentions a syndactylic

An unusual feature was that the recurrent attack occurred inside the sac of a post-operative ventral hernia, and this complication helped to cause some difficulty in pre-operative diagnosis.

The absence of tenderness over the hernial sac and of rise of temperature pointed to the condition being one of incarcerated hernia rather than to its being an acute gangrenous perforated appendix, with the presence of pus.

I am indebted to Mr. Lionel Stretton for permission to publish these cases.

family of thirty-nine members, fourteen of whom had the deformity. Sverdrup⁴ describes it in six generations of a Norwegian family. Epstein reports it occurring in five generations, transmission occurring in male and female.

A valuable monograph² by Messrs. Lewis and Embleton gives a very exhaustive investigation of the members of one family in which there is a record of forty-four members having deformities of the hand and foot. Douglas P. Murphy⁵ has made a study of the deformity in five successive generations.

In the Johns Hopkins Hospital Bulletin⁶ of 1927 a case was reported of combined acrocephaly and syndactyly occurring in mother and daughter. The two cases reported support the view expressed by Park and Powers that the combined malformations are evidences of germ-plasm defects rather than the results of externally operating causes.

CASES DESCRIBED.

I have been able to obtain full details from the living members of this family which I am about to describe. Unfortunately I have not been able to trace the great-grandmother, but she was definitely known to have deformed hands.

The grandmother, aged 65, has syndactyly of both hands and has never undergone operation for the deformity. From the accompanying photograph (Fig. 1) it will be seen that there is complete syndactyly of the left hand, with that rare occurrence the fusion of the thumb and index finger. The dorsal surface of the hand is very misshapen on the ulnar side, with marked bosses over the interphalangeal joints. The nails are very large and there is a slight division in them, showing attempts at separate nail formation. The right hand has complete syndactyly, but the thumb is separated in a rudimentary form (Fig. 2). There are less fingernails on the right hand, and they are marked by grooves, showing attempts at separation. In spite of these deformities she is quite dexterous, and can manage all simple household work.

The mother, Mrs. X, aged 33, has syndactyly of both hands. She came under the care of Mr. Openshaw at the London Hospital in 1897, when she was 3 years old, and was successfully operated upon. She has one sister who is similarly affected, and three other sisters who are quite normal in this respect; there was also a stillborn brother who was said to have had the prevalent defect. Originally in both hands the fingers were enclosed in a web of skin.

Both Mrs. X and her sister had six sets of digital bones on the right, but the mother and one of the other child's sixth set were more complete than Mrs. X's, whose sixth finger consisted of a small rudimentary process of the other side of the hand. The digital bones otherwise appeared quite complete, and the movement suggested that the muscles were normally attached. At the distal part of the web of skin was a ridge of nail-like substance representing the normal nails. The thumb was partially separate on each hand.

In a period of three years fifteen operations were performed, and as a result she now has five very useful and completely functioning digits on each hand. She is able to wear a wedding