

A CASE OF CHRONIC FAMILY JAUNDICE.

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CASE HISTORY. Miss A. H., aged twenty-four years, an office clerk, complained of her abnormal color and of severe attacks of abdominal colic. Her mother has always had jaundice, and large numbers of gall-stones were removed at operation. Her grandfather, her uncle, and her aunt, all on the mother's side, were said to have suffered from a similar condition. Her brother is apparently well.

There is nothing definite in the patient's personal or past history until she arrived at ten years of age. At that time she became "terribly" jaundiced, had fever and severe pains in the abdomen, and the abdomen increased in size. An enlargement of the liver and spleen was noted. She was confined to bed for six or seven months. Although she improved after that, her jaundice never entirely cleared up, and there were distinct exacerbations at intervals. Three years ago she had an attack of appendicitis, and her appendix and right ovary (reason not determined) were removed.

About a year ago the patient began to have attacks of pains in the epigastrium with radiation to the right shoulder. There was no nausea, but she forced herself to vomit, seeming to get some relief in this manner. She has had three severe colic attacks during this interval, the jaundice increasing after each; the urine was also observed to grow darker. The patient had no chills, but felt "clammy" during each exacerbation. She observed no regular relation between the onset of pain and the ingestion of food, but the pains seemed worse when she was nervous. There was very slight digestive disturbance, a little sour stomach lately, some regurgitation at times, and bloating. The bowels were costive. During the summer the patient had frequent nose-bleeds, but not lately. There were no symptoms (with the possible exception of slight dizziness) which could be attributed to the cardiovascular system.

Except for the occasional pains and the constant discoloration of skin the patient feels perfectly comfortable, and is happy.

Physical Examination. The patient is well developed and well nourished, active, and of a cheerful disposition. The conjunctivae are distinctly icteric, the skin of the body moderately so, the color of the face, especially after exposure to the cold air, may be described as of a delicate tea-rose shade (icterus plus anemia).

The heart offers some points of interest. Its width by percussion at the level of the fifth space is 12 cm. This is over 46 per cent. of the total chest width at that level (normal value being $41\frac{2}{3}$ per cent.). The heart is therefore distinctly enlarged. Fluoroscopically this finding is confirmed and the organ appears to be of a "mixed" configuration. On auscultation there is a loud, rough, systolic murmur at the pulmonic area. At the junction of the



FIG. 1.—Radiograph of the heart enlarged both to right and left.

fourth rib and the sternum another loud systolic murmur, softer in quality, is audible. This latter is transmitted downward toward the apex. The pulse is of good quality, 76, and regular. These findings were rather a surprise, as the patient presented no subjective evidence of cardiac involvement. Moreover, anemia could hardly be invoked to explain the entire picture. A possible congenital defect was thought of.

The blood-pressure was 100 mm. systolic and 55 mm. diastolic. Hypotension has apparently not been mentioned as characteristic of the disease, although most reported figures are low.

The liver edge could not be felt, but the left lobe percussed 6.5 cm. out from the spine by the Grocco method. There was tenderness at the junction of the right midclavicular line and the costal margin, as well as some resistance just below this region. The spleen was large, firm, smooth, and not tender. Its lower pole reached 8 cm. below the costal margin in the midclavicular line, its upper border to the eighth space, 13 cm. above the rib margin

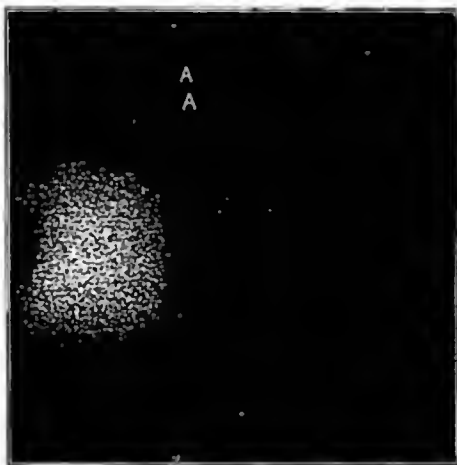


FIG. 2.—Radiograph of gall-bladder region showing two gall-stones: A, gall-stones.

in the mid-axillary line, and its anterior (right) border to just within the mid-line. Its total oblique (greatest) diameter was 16 cm.

Laboratory Findings. Blood: The coagulation time by the vein puncture method was five minutes (normal control eight minutes). The hemoglobin percentage (Sahli) was 55, 53, 65, 56 on four occasions. The total red cells numbered 2,400,000, 2,700,000, and 2,540,000, the color index being respectively 1, 1, and 0.98. Hemolysis of the erythrocytes (whole blood was used) began at 0.5 per cent. NaCl on two occasions and at 0.55 in another, the process being complete at 0.425 and 0.45 respectively. Normal bloods began to

hemolyze at 0.425 and 0.45, and were completely laked at 0.325. The blood serum was positive for bile pigment (Hedenius's method). The Widal-Abrami-Brulé auto-agglutination test was negative. There was a slight though positive increase (as compared with a normal control) in the relative number of red cells showing vital staining. The size of the erythrocytes was diminished. The leukocyte counts were 3800 with 62 per cent. polymorphs and 4500 with 66 per cent. polymorphs. The Wassermann test was negative.

Urine: High colored; specific gravity 1013 to 1018; slightly acid; never bile; urobilin always present (Schlesinger test); also trace of albumin, otherwise negative.

Stool: Normal in color.

Test meal: Fasting stomach empty; Ewald meal: contents well digested, free HCl 28, total 48.

Röntgen-ray Examination. Esophagus negative, stomach vertical, fair tone, lower pole 4 inches below navel; cap normal; empty of Rieder meal within six hours. Intestines fill and empty normally.

Plate of gall-bladder region shows four small shadows, two below and two upon the same level as the twelfth rib. Shape and position characteristic of gall-stones.

Discussion. The question of therapeutics is of interest in this case. At present the patient is receiving iron, is free from pain, and operative treatment is not regarded as urgent.

In their excellent study of chronic family jaundice (1910), in which they introduced the subject to American clinicians, Tileston and Griffin¹ had the following to say concerning the nature of abdominal colic in this disease:

"Attacks of abdominal pain resembling biliary colic have been observed in a large proportion of cases, and have been often supposed to be caused, in some mysterious way, by the disease itself. In two of our cases, however, gall-stones were removed at operation, after which the attacks ceased, and in both our autopsies stones were found in the gall-bladder, so that we have no hesitation in declaring that these attacks are due to gall-stones. A striking feature is the frequent onset of the colic at the time of puberty, a period at which ordinary cholelithiasis is seldom met with."

And further, under the heading of treatment they remark:

"It should be realized that attacks of abdominal pain are not due to the disease *per se*, but to a complication with gall-stones, and the patient should be given the benefit of modern surgical treatment, which has been neglected heretofore in all cases except in two of our series."

Since the above was written, evidence has been adduced tending

¹ Chronic Family Jaundice, *Am. Jour. Med. Sc.*, 1910, *cxix*, 847.

to show that colic may be present without stones (case of Thayer and Morris²) in which no stones were found at operation, also two cases quoted by Elliot and Kanavel³ and that the removal of the stones may not produce absolute relief from pain (case of Richards and Johnson⁴); relief for one year, then a mild attack). Perhaps the existence of intrahepatic calculi may explain these apparent discrepancies.

Finally, a word as to the cardiac condition. In a review of the literature from this stand-point, we found no mention of a similar condition, until we came to the report of Hiehens⁵ who recently presented before the Royal Society of Medicine three cases, and mentioned a fourth, of familial jaundice (in one family), each of whom showed a loud systolic murmur heard best in the third space, and transmitted upward and to the apex. This author also suggested a possible congenital origin for the condition.

Roentgenograms of the heart and stones are appended.

Since writing the above, the patient had another severe attack of gall-stone colic, and became deeply jaundiced. She was advised to have the stones removed. She entered St. Joseph's Hospital, and on January 3, 1916, Dr. Frederick Flaherty removed two fair-sized stones and a number of small ones. The ducts were free.

AUTOSEROBACTERINS.

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In the present communication we propose to outline in brief the method of modifying sensitization of vaccines as carried out at the Nicholas Senn Hospital. A full discussion of this phase and of vaccines in general has been given in another place.¹

Besredka,² in 1902, introduced sensitized vaccines into medicine. By this is meant that a suspension of bacteria (dead or alive) is mixed with serum from an immune animal and allowed to macerate

¹ Two Cases of Congenital Hemolytic Jaundice with Splenomegaly, Johns Hopkins Hosp. Bull., 1911, xxii, 85.

² Splenectomy for Hemolytic Icterus, Surg., Gynec. and Obstet., 1915, xxi, 21.

³ Study of a Case of Coagenital Hemolytic Jaundice, Jour. Am. Med. Assn., 1913, lxi, 1586.

⁴ Cases of Familial Jaundice, Proc. Roy. Soc. Med., 1913, ix. Section of Studies of Diseases of Children, 197.

⁵ Med. Record, April 29, 1916.

⁶ Comp. rend. Acad. des sci., 1902, vol. c, xxxiv.