semilunar cartilage be recorded in a similar surgical crescendo?

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TREATMENT OF FAMILIAL ACHOLURIC JAUNDICE

BY

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Familial acholuric jaundice, first described by Minkowski in 1900, is now recognized to be of widespread though not of common occurrence. Its peculiar characteristics have attracted an amount of attention in medical literature which the relative infrequency of its incidence scarcely justifies. Comprehensive reviews by Tileston,¹ Dawson,² and Cheney and Cheney,³ amongst others have made the general features of the disease well known. The pathogenesis, however, remains obscure in spite of Haden's⁴ illuminating work on the fundamental abnormality of the erythrocyte. These aspects of the subject will not, therefore, be dealt with in this paper, which will confine itself to a brief review of the results of splenectomy, and a discussion of some problems of treatment about which general agreement has not yet been reached.

The Clinical Material

Familial acholuric jaundice appears to be of relatively common occurrence in Victoria; during the past fifteen years I have seen thirty-three patients suffering from the disease. Seventeen of the cases have occurred amongst the members of the Hazel family, which has already been the subject of several communications. Springthorpe⁵ in 1904 described the clinical features presented by six of its affected members, on two of whom splenectomy had been performed successfully by Stirling. In 1908 he⁶ recorded the subsequent history of these patients, and drew attention to the lasting benefit which had followed removal of the spleen. Unfortunately Springthorpe used the term "splenic anaemia" in describing his cases; this error in nomenclature has prevented his pioneer work from receiving the recognition it deserves. In 1922 I brought the details of the family history as far as possible up to date.⁷ Since then some additional information has been obtained, and this is incorporated in the accompanying family tree.

Clinical evidence sufficient for diagnosis has been obtained in thirty-four individuals; of twenty-six who have come under my care seventeen have been shown to present the typical features of acholuric jaundice, and ten of them have been treated by splenectomy. In addition, sixteen other patients suffering from familial acholuric jaundice, representing eight affected families, have been seen in private practice, and ten of them have had their spleens removed. The series of personally observed cases on which this paper is based consists, then,

of thirty-three patients, twenty of whom have been subjected to splenectomy.

Results of Splenectomy

The results of splenectomy in this series bear out the generally accepted view that the operation brings about complete and lasting freedom from symptoms. One point which has not been sufficiently emphasized in the literature is the rapidity with which the more seriously affected patients improve after operation. The jaundice disappears almost immediately, and the anaemia improves so rapidly that within six weeks a bed-ridden child may be well enough to return to school. Because of the dramatic nature of the improvement pre-operative transfusion is seldom needed, and splenectomy may be undertaken without hesitation, even in patients who appear almost desperately ill.

The operative mortality is small. In this series no deaths have occurred from causes in any way related to the operation. Two patients have died, one from pulmonary tuberculosis three years after splenectomy, and one from lobar pneumonia eighteen years after. Experience in other parts of the world also shows that the risks of operation are slight. Pemberton⁸ reports from the Mayo Clinic 118 cases with four deaths in hospital, Vaughan⁹ nine cases with one death, Herfarth¹⁰ 176 with six deaths, and Beckman and Jäderholm¹¹ had eighteen cases with no primary mortality.

Operative morbidity also has been negligible in this series. One child, aged 10 years, and one adult, 42 years of age, developed purulent effusions in the left pleural cavity; both recovered rapidly after aspiration. The technical difficulties imposed by the presence of gall-stones will be referred to later. In several instances among the adult patients dense peri-splenic adhesions have rendered the operation tedious. But in general it may be said that few of the cases have given rise to any real anxiety.

The remote results of splenectomy have been equally satisfactory. Of the two patients operated on by Stirling in 1904 one bore four children and remained free from symptoms for eighteen years until her death from lobar pneumonia, and the other, who has had seven children, is still in good health. All the other patients who have been surgically treated have now remained well for a period sufficiently long to bear out the general statement that the relief afforded by splenectomy is permanent.

The Indications for Splenectomy

It is generally agreed that the administration of liver or iron, and the use of x rays or radium, has little or no effect on acholuric jaundice. The management of the individual case turns, therefore, on whether the spleen

should be removed, and if so at what stage. Despite the successful results of the operation there are differences of opinion about the indications for its performance. Influenced, no doubt, by Chauffard's early dictum that patients suffering from the familial type of acholuric jaundice are more jaundiced than sick, most authorities state that splenectomy should not be advised unless the symptoms are so severe as to cause definite incapacity. This attitude is justified by the fact that the disease, even if it is untreated, is often compatible with long and useful life. On the other hand, the occasional occurrence of severe, and sometimes fatal, haemolytic crises, which may occur quite unexpectedly, and the greater difficulty and danger of the operation in older patients support the opposing view that splenectomy should never be long

and increased fragility of the red cells was present, haemolysis commencing in 0.525 per cent. saline (normal control 0.425 per cent.). The blood findings were: red blood cells 2,190,000 per c.mm.; haemoglobin, 35 per cent. (Sahli); white blood cells, 10,300 per c.mm. Film: gross microcytosis; polychromatic cells very numerous and of intense blueness; many normoblasts and an occasional myelocyte seen.

Splenectomy was advised and pre-operative preparation by transfusion was deemed unnecessary. Mr. Alan Newton removed at the one operation the spleen and a chronically inflamed gall-bladder containing six pigment stones. Convalescence was rapid and uneventful, and the patient has since enjoyed good health.

In this case the prognostic significance of the family history afforded an indication for splenectomy which

HAZEL FAMILY TREE.—Figures inside circles denote age of patient. Roman numerals denote cases personally observed.

delayed once the diagnosis has been made. This divergence of opinion is doubtless due to the occurrence of wide variations in the severity of familial acholuric jaundice. It is, however, not sufficiently recognized that, generally speaking, the disease follows much the same course in the individual members of each affected family. In other words, the variations in severity are familial rather than individual. In the Hazel family, for instance, although the disease has caused much incapacity and invalidism amongst its members, not one has actually died of its effects, and I have therefore felt justified in advising splenectomy only when the symptoms have become sufficiently severe to interfere with the patient's economic and social activities.

In other families acholuric jaundice occurs in a much graver form, as the following case history shows.

CASE REPORT

A female, aged 16 years, was the only survivor of a small family. Her father had been accidentally killed. Her mother had died at 31 and her only brother at 16 years of age from severe anaemia which had been regarded as pernicious in type. When I saw her, in 1922, she presented the classical features of acholuric jaundice in a severe form, and there can be no doubt that it was this disease which caused the deaths of her mother and brother. She was grossly jaundiced,

could not be disregarded. Dawson² has described a similar case, "B. Tope," who was less ill than my patient, but whose family history was equally ominous. Of her he remarks: "Can there be any doubt that this child, however well she appears, should have her spleen removed?" With this attitude I heartily concur, and I would urge that in every case of familial acholuric jaundice the family history, as well as the patient's general health, should be considered in arriving at a decision for or against splenectomy.

Treatment of Associated Pigment Stones

In seven of the twenty patients treated surgically in this series pigment gall-stones have been found, but in only one of them had a mistaken diagnosis led to a previous operation on the biliary tract. These figures are lower than those recorded by other observers. For instance, Pemberton³ reports that 68.6 per cent. of patients at the Mayo Clinic showed conclusive evidence of disease of the gall-bladder, with and without stones, and that 20 per cent. had previously been subjected to operations on the biliary tract, presumably without knowledge of the presence of the primary disease. Vaughan⁴ found stones present in six of seven patients whose gall-bladders she examined. It would seem that pigment

stones occur more frequently than my own figures indicate.

There is, as yet, no general agreement about the best method of dealing with these stones. Vaughan perhaps expresses the accepted view when she says: "At the time of operation it is essential to remember the frequency of gall-stones, and if possible to remove the gall-bladder at the same time." This attitude appears illogical. The biliary stones are not due to primary disease of the gall-bladder; if cholecystitis is present, as in the case cited above, it must be regarded either as an accidental complication or possibly as the result of long-continued irritation by pigment stones. The formation of stones results from the deposition of a portion of the large amount of pigment with which the liver cells are called on to deal. If excessive haemolysis is terminated by splenectomy the gall-bladder has only to be emptied, not removed, and recurrence of stones need not be feared. The correctness of this attitude is supported by the results obtained in my own cases.

FIVE CASES

In three patients, operated on more than ten years ago, cholecystotomy and removal of stones was done at the same time as the splenectomy; in none of them has any evidence of biliary disease since manifested itself.

In another patient, a boy 12 years of age, Mr. Newton contented himself with crushing the friable stones digitally, without opening the gall-bladder; there has been no recurrence.

The last case, a man aged 60, is the only one of this series who had previously been operated on for gall-stones; cholecystotomy had been performed twelve years before by another surgeon and a number of stones removed. When Mr. Newton reopened the abdomen he felt a large pigment stone in the gall-bladder. After he had removed the spleen he opened the gall-bladder only to find that the stone had disappeared; doubtless it had been crushed during the manipulation of the viscus. The incision in the gall-bladder was closed with a purse-string suture. This was five and a half years ago, and there has been no recurrence.

Practical experience therefore supports the conclusion reached on theoretical grounds that the proper treatment of the pigment stones so often encountered in familial acholuric jaundice is by splenectomy and by simple emptying of the gall-bladder. This can usually be accomplished by crushing the stones digitally, but cholecystotomy may be necessary. The gall-bladder should be removed only if definite evidence of cholecystitis is present.

Summary

The results of splenectomy in twenty patients suffering from familial acholuric jaundice are reviewed briefly. There has been no operative mortality, and the operative morbidity has been negligible.

Variations in the severity of the disease are familial rather than individual. The family history, as well as the clinical findings in the individual patient, should be considered in determining the advisability of splenectomy.

When biliary pigment stones are present the gall-bladder should be emptied by crushing the stones digitally or by cholecystotomy. Cholecystectomy should be performed only when evidences of cholecystitis are present.

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TEST-MEAL FINDINGS AND BILIARY DISEASE

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In carrying out routine Rehfuß test meals it was noticed some time ago that certain curves had much greater differences between "free" and "total" acids than others. Further inquiry showed that these abnormal ones were suspected of having lesions of the gall-bladder or biliary tract. An effort was made to collect curves from all such cases available; up to the time of writing fifty-one have been examined, and of these twenty-two have been operated on and three have been x-rayed only. These twenty-five cases in which the diagnosis has been established, either by operation or by positive x-ray findings, will now be considered.

Standard Technique

At 9 p.m. the night before the test four small charcoal biscuits are given with a cup of tea, and nothing further is taken for twelve hours. The test meal consists of a gruel prepared by simmering a quart of water, with two tablespoonfuls of medium oatmeal, down to one pint and straining through muslin. The volume is then corrected to a pint with water. A narrow, rubber-ended Ryle's tube is passed, to the second mark, at 9 a.m., and the resting juice removed; two ounces of distilled water are injected into the stomach and removed; and the test meal is given with instructions to drink it all within five minutes, if at all possible. Specimens of from 5 to 10 c.cm. are then withdrawn at fifteen-minute intervals for three hours. All specimens are filtered to ensure sharp end-points to the titrations. One c.cm. of the filtrate is taken, with a pipette graduated in 1/100 c.cm., and is titrated with N/100 NaOH, using Töpfer's reagent for the free HCl and phenolphthalein for the total acids. A graph is then constructed showing the "free" and "total" acids as cubic centimetres per cent. of N/10 HCl. A further curve shows the differences between "free" and "total" acids in the same units, but for the sake of clarity the scale of this difference curve is five times that used for the "free" and "total" acids.

Test-meal Findings

In normal and non-biliary system cases the difference curve rarely rises over 12 c.cm. (Fig. 1).

FIG. 1.—A normal test-meal finding, showing the difference curve between "free" and "total" acids.

The broken line represents free HCl; the dotted line total acidity; and the continuous line the difference between "free" and "total" acids.

In cholelithiasis and cholecystitis and infective conditions of the biliary system the difference curve rises as high as 50 c.cm., and in the series under review is always