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# FAMILY TENDENCY TO HYPERTROPHIC CIRRHOSIS OF THE LIVER (HANOT'S DISEASE), BY JAMES FINLAYSON, M.D., LL.D.

# FAMILY TENDENCY TO HYPERTROPHIC CIRRHOSIS OF THE LIVER (HANOT'S DISEASE).

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THE occurrence of great enlargement of the liver, associated with jaundice, has long been known; under the name of hypertrophic cirrhosis it is now generally recognized, especially since the teaching of Hanot. With this liver affection we often have enlargement of the spleen, without leukaemia; abdominal dropsy may occur as a complication, but is rare if the comparison is with the regular atrophic cirrhosis. As in the case of the last-named disease, the injurious influence of alcohol may be traced as an etiological factor; but in very many cases of hypertrophic cirrhosis this may be excluded. Of special interest, on this and other accounts, is the study of this form of disease in children, and many cases have now been published in young subjects.

In 1898 I had under my notice a very typical illustration of this disease in a young lad, then 18 years old, a worker in the coal pits; he had been affected for two years at least, and he was not open to the suspicion of alcoholism. A prolonged study of his case, while resident in the Western Infirmary (May 31st, 1898), failed to reveal any new feature. He had very troublesome itching of the skin, which varied much in severity from time to time; and he had had repeated attacks of epistaxis, some of which we witnessed, but no haemorrhages from the stomach or bowels had occurred. The jaundiced

tint was distinct both in the skin and conjunctivae although not extreme; the urine was dark in colour and showed a slight reaction of bile pigment on testing with iodine, very indistinctly with nitric acid, or not at all. The motions were sometimes clay-coloured, at other times natural, without any corresponding variation in the jaundiced tint; no dropsy; enormous enlargement of the liver, which felt hard and smooth; no distinct enlargement of the spleen could be made out, a very definite tumour in the left hypochondrium being regarded as the left lobe of the liver, as it seemed continuous with that organ; no glandular enlargements anywhere.

His previous history was that of frequent attacks of feebleness and of bad health, with some little abscesses, but nothing either in himself or in the family, which was a moderately large one, appeared to suggest syphilis. He seemed, for his age, somewhat under-developed. Under treatment he improved a little, but his condition was not materially altered when he left; indeed, he seemed very much as before when seen again at the Infirmary a year later (May, 1899). He then weighed 7 st. 1 lb., much the same as the year before.

On his return, however, a new point of great importance emerged. His medical attendant, Dr. Muir of Bellshill, reported that the patient's sister, two years older, was similarly affected, and they were sent in together for comparison. The sister presented a striking family resemblance to her brother. She too was under-developed, and her menses had never appeared, although she was 21 years old. She also had suffered from feeble health at least since she was 14 years: her colour was exactly the same as her brother's; there was the same slight reaction of bile pigment in the urine. She too had been troubled with itching of the skin, although not in such a marked form. She also had repeatedly lost blood from the nose. No dropsy existed, and there had been no haemorrhages from the stomach or bowels. In her the liver was greatly enlarged—not quite so extremely as in her brother's case; but in her the spleen was felt to be greatly enlarged. Indeed, on examining the

abdomen, the splenic tumour was more striking than the enlarged liver. No glandular swellings were detected, and there was no alcoholic history. She weighed 7 st. 11 lbs.

An examination of the blood was made by Dr. Carstairs Douglas during the lad's residence in 1898, and again when he returned with his sister. No increase of the white blood corpuscles existed in either case; indeed, some of the estimates made them less than the normal. The number of the red blood corpuscles was about the normal in both patients, and the haemoglobin was not much below par.

This discovery of a second member in the family being affected led Dr. Muir to examine some of the other members of the same family, and he reported that a third member, a younger brother, was also found to have a large liver and spleen. Dr. Muir supplied the following notes of this lad:— Age, 15 years; affected for 18 months; jaundice slight; itching occasional; bleeding from nose sometimes; liver large, extending 3 inches below the ribs; spleen enlarged, extending 3½ inches below the ribs; no history of alcohol. Another brother, still younger, seems a little yellow, but no enlargement of liver or spleen can be detected in his case.

Three members of a family similarly affected with a condition relatively rare in early life led to a search into the literature of cirrhosis of the liver, especially as regards young subjects or family proclivity.

The little memoir of Hanot does not indicate any experience of family tendency in this disease, although he refers to the subjects being weakly and poorly developed (p. 99). In Osler's *Practice of Medicine*, third edition, 1898, p. 574, the writer states, under the heading of "Hypertrophic Cirrhosis," that two of the cases he had seen were in brothers. In the *International Journal of the Medical Sciences* (Oct., 1887, p. 350), in an article on "Hepatic Cirrhosis in Children," Dr. R. P. Howard gives the case of a girl, 9 years old in 1878, affected with a large liver and spleen, with a history of epistaxis, slight pyrexia, and latterly ascites. Her brother became icteric in 1884; he also had enlargement of the liver and spleen, and he also had slight pyrexia and ascites.

Neither of them had leukaemia. Both died, and in both the livers are described, post-mortem, as large and granular.

In the *British Medical Journal* for April 23rd, 1892, p. 858, Mr. Jollye reports two cases in children, a brother and sister, who died with ascites due to atrophic cirrhosis; although a post-mortem examination was obtained only in one case, he presumed the condition was similar in the other. He also quotes Dr. Ormerod's paper, which falls to be noticed hereafter.

The existence of a family tendency to hypertrophic cirrhosis is obviously important, although its significance is still obscure. Of course, if due to syphilis, the explanation might be clear; or, if due to transmission from a drunken parent (see Hanot, p. 99), the disease might well occur in various members of the family. Both of these causes seem fairly to be excluded in the family here referred to.

The resemblance of hypertrophic cirrhosis to what is called "biliary cirrhosis" seems very close. Further, in hypertrophic cirrhosis, we have a disease characterized by jaundice, one occurring in early life, and specially implicating certain families; it may be well, therefore, to consider some of the features of congenital malformation of the bile ducts where these peculiarities are also found.

The occurrence of biliary cirrhosis, with enlargement of the liver and often with enlarged spleen also, is well known in cases of congenital obliteration of the bile ducts. "It seems probable that biliary cirrhosis always occurs if the child lives long enough" (Dr. John Thomson, *Congenital Obliteration of the Bile Ducts*, Edin., 1892, p. 29). Dr. Thomson also says, of such congenital defects, "there is evidently a very remarkable tendency for the disease to occur in more than one child of the same parents" (p. 13). In his tabulation of cases we find one (No. 22) where one twin died with obliteration of the duct, and the other, although jaundiced, recovered. Other cases (No. 9 and No. 33) in his table show recoveries from icterus neonatorum in families where fatal defects of the ducts had occurred. Further, cases are given in table No. 4, where, with similar symptoms proving fatal and in families

affected with this grave form of icterus, the ducts were found to be still pervious. From all this we may infer a great degree of variation as to the severity or extent and also as to the duration of the affection of the bile ducts, or the period in intra-uterine life when the disorder appears. It is also important to notice that Dr. Thomson shows that this defect is sometimes associated with other manifestations of developmental disorder. In my cases of hypertrophic cirrhosis the brother and sister were evidently under-developed, and in particular the girl, although 21 years old, had never menstruated. The remark already quoted from Hanot (p. 99), as to the disease occurring in badly-developed subjects, may be recalled in this connection.

In Dr. Ormerod's paper (*St. Bartholomew's Hospital Reports*, vol. xxvi., 1896) reference is made to the occurrence of cirrhosis of the liver in a young subject associated with a family tendency to some peculiar nervous disorder; but, in the absence of details, not yet published, it is difficult to estimate the value of this group of cases for our present purpose. In the same paper he quotes the experience of Prof. Homén (*Neurologisches Centralblatt*, 1890, p. 514), where two members of a family had this combination of well-marked cirrhosis of the liver and some nervous lesion (in the lenticular nucleus) when examined after death; a third member of the family seemed to be similarly affected. The illnesses ran a chronic course of six or seven years, and proved fatal at the age of 26 and 19 years respectively.

In the case of family cirrhosis we may suppose that a developmental defect occurs in the liver in certain members of the family; the mere fact of the common bile duct being found pervious at death in cases of hypertrophic cirrhosis (Hanot, p. 132) does not exclude the notion of a possible defect or peculiarity in the ducts determining a biliary cirrhosis. We know that very slight obstructions interfere with the flow of bile, and some defect in the lumen of the ducts within the liver itself might lead to disorder. We have instructive analogies in the congenital defect of the bile ducts, already referred to, viz. cases with similar symptoms occurring at

birth, proving fatal, with ducts still pervious; and, on the other hand, cases of recovery in jaundiced infants whose brothers or sisters died with the developmental defect of the ducts. Still further, there is a case of some importance from this point of view communicated by Dr. Fred. J. Smith to the Pathological Society of London (*Pathological Transactions*, vol. xli., p. 154, 1890), where an infant recovered from an attack of icterus neonatorum after a fortnight, but became the victim of hypertrophic cirrhosis, with greatly enlarged spleen, without dropsy, dying at the age of 4½ years. To be compared with this is the family described by Pearson in Underwood's treatise on *Diseases of Children*, 4th ed., 1799 (see Thomson's Table IV., No. 1), where nine children in the family died under a month old, from jaundice, and the tenth died from jaundice also, when six years old; in the post-mortem examination of the eleventh child in this family, who also died of jaundice on the ninth day, the ducts were found to be pervious and the gall bladder full of bile.

Such ideas arise in connection with my three cases of hypertrophic cirrhosis in one family, when compared with other reports showing a family tendency to this disease. It must be remembered, however, that the cases called by this name constitute still a very mixed group. Indeed, it may be, as contended by some, that these enlarged livers might in time become small and granular. The records of Dr Howard's cases, of a brother and a sister, showed large livers in both, but in both the surface was granular.

If attention is called to the question of family tendency in hypertrophic cirrhosis, especially in early life, some further light may arise when more cases are definitely investigated; for it is worthy of notice that in my first case, although the family history was inquired into, nothing of this kind was elicited, and it was only from Dr. Muir's personal knowledge of the family that this important peculiarity was noticed in the second case, and searched for in the third.

## REFERENCES.

THE little book of Hanot, *La cirrhose hypertrophique avec ictère chronique*, Paris, 1892, gives a full account of the whole subject, with historical references, including his own thesis, *Sur une forme de cirrhose hypertrophique*, Paris, 1875. There is an elaborate review of the subject, up till 1877, in *The British and Foreign Medico-Chirurgical Review*, July, 1877, "Hypertrophic Cirrhosis of the Liver." An important tabulation of cases of "Cirrhosis of the Liver in Childhood," or early age, is given by W. A. Edwards in the *Archives of Pediatrics* for July, 1890, vol. vii., p. 502, where he also contributes a case of the hypertrophic form. Dr. Mary Putnam Jacobi has an interesting article in the *Archives of Pediatrics*, May, 1889, vol. vi., p. 273, "Case of Cirrhosis of the Liver with Splenic Tumour" (girl 10 years old); in this paper she quotes various authors, amongst others, Laure et Honorat, "Étude sur la cirrhose infantile," *Rev. mens. d. mal. de l'enfance*, t. v., Paris, 1887. (I quote the reference from the *Index Medicus* as I have not seen the original.) Dr. R. Palmer Howard, "On Hepatic Cirrhosis in Children," in *International Journal of the Medical Sciences*, October, 1887, p. 350 (a sister and brother). F. W. Jollye, "Hepatic Cirrhosis occurring in Two Children of the same Family," *British Medical Journal*, April 23rd, 1892, p. 858. Ormerod, "Cirrhosis of the Liver in a Boy," *St. Bartholomew's Hospital Reports*, vol. xxvi., 1890, p. 57; this paper has several points of interest in connection with the question of family tendency, and in particular he gives a reference to Prof. Homén, *Neurologisches Centralblatt*, Bd. ix., 1890, p. 514, referred to above, where two or probably three in a family were affected with cirrhosis of the liver and some rare nervous disease. The title in *Index Medicus* is "Eine eigenthümliche Familienkrankheit unter der Form einer progressiven Dementia mit besonderem anatomischen Befund." Osler's *Practice of Medicine*, 3rd edition, Edinburgh, 1898, p. 574 (two brothers with hypertrophic cirrhosis). The literature of hypertrophic cirrhosis of the liver is now very considerable, and that of cirrhosis of the liver in childhood is also large, as may be found on consulting the *Index-Catalogue* of the Washington Library, and the *Index Medicus*; many of the cases are tabulated in papers quoted above.