EDWARDS (Wm A.)

Cirrhosis of the liver

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CIRRHOSIS OF THE LIVER IN CHILDHOOD.

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WITH MICROSCOPIC REPORT.

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In the present communication, it is our desire to further study the case which formed the basis of a paper on the subject of hepatic cirrhosis in children that appeared in the ARCHIVES OF PEDIATRICS, in July, 1890. We desire, also, to present the results of the post-mortem examination in this case, together with the microscopic studies, and the photo-micrographs of Dr. Gray, and more particularly to consider the etiological factors in the production of cirrhosis of the liver in childhood. The child came under my observation in June, 1888, and died in my private hospital September 24th, 1890, age 12 years and 4 months. Two months after, the clinical notes were recorded in the ARCHIVES. The disease pursued about the usual course as seen in the adult. Gastric disturbances, coated tongue and foul breath, abdominal pain, slightly augmented by pressure, occasional nose bleed, stigmata composed of collections of dilated minute venules, constipation, alternating with diarrhœa, headache, dry harsh skin, some side pain, marked jaundice, lassitude, languor, and drowsiness. Thirty days before

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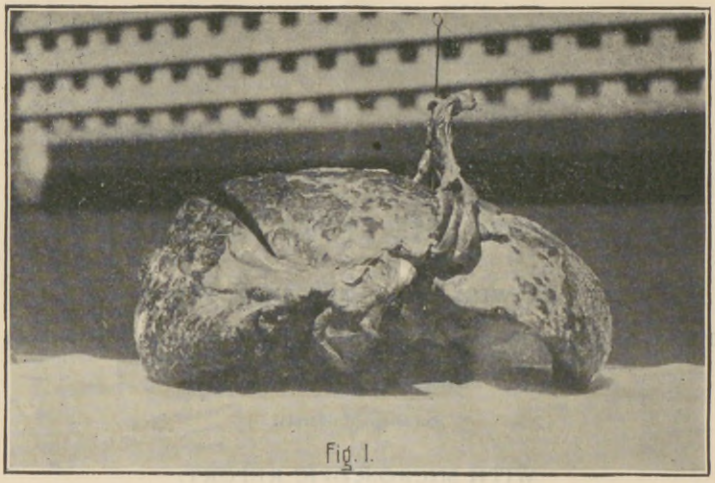

FIGURE 1. SUPERIOR SURFACE OF LIVER; PIN THROUGH SUSPENSORY LIGAMENT. ANTERIOR EDGE SHOWS REMARKABLE CONNECTIVE TISSUE FORMATION AND LOBULATION.

FIGURE 2. UNDER SURFACE OF THE LIVER, SHOWING LOBES, GALL BLADDER, AND THE EXTRAORDINARY LOBULATION OF THE PARENCHYMA.

death, ascites arose, in twenty days the belly became enormously distended with serum. The legs were œdematous, the veins of the breast and abdomen were greatly distended. The pulse was 120, respiration 30, temperature 38° C. The urine contained a small amount of albumin and a few granular tube casts. Total quantity passed in twenty-four hours, 500 c. c.

On September 13th, 1890, assisted by Dr. J. P. LeFevre, I performed paracentesis abdominis, and withdrew four and one-half litres of clear fluid. For twenty-four hours, there was a free drainage of clear serum from the puncture, the next day it decreased to almost nothing.

The child became drowsy and complained of abdominal distress and pain. The original puncture in the abdominal wall was enlarged, and a rubber drain inserted, at once 1,000 c. c. of fluid drained away. This fluid however, was no longer clear serum, but contained some pus. Slight improvement was noted in the child's general condition; the temperature remained lower, the pulse below 100, and the respiration below 20. This gain was, however, of short duration. The urine became scant, high colored, and bile stained, and was voided with difficulty. The child grew progressively weaker and weaker, hiccough became worse, the temperature subnormal, and the pulse extremely irregular. The urinary excretion was almost supressed, and finally, on the evening of the 23d, convulsions arose, which persisted until death on the 24th. These convulsions were most severe, clonic in nature, and lasted for fifteen or twenty minutes, with only short intervals between. Death occurred in one of these spasms.

Autopsy, six hours after death, assisted by Drs. LeFevre and T. A. Davis. Child much emaciated; several petechial spots over thighs, legs, and chest. Thorax: heart small; cavities dilated and contained clots; valves, normal; aorta, healthy; muscles, slightly fibroid; lungs and pleuræ, healthy. Abdomen: peritoneum, thickened, and venous channels greatly dilated. Stomach: mucous membrane thickened and slightly eschymotic; spleen presented a

normal appearance, with the exception of intense congestion, and consequent increase in size.

The gross appearance of the liver is represented in the accompaning photographs, kindly taken by Dr. F. H. Mead, of San Diego. It weighs 665 grams, and is a remarkable example of atrophic cirrhosis. Its microscopic changes are most graphically and accurately presented in the photo-micrographs of Dr. Gray. The uncommon increase in inter- and intra-lobular connective tissue is well shown.

The gall bladder was comparatively normal, its mucous membrane slightly thickened, contained a small amount of bile, but no calculi.

The kidneys were large, swollen, congested, and hyperæmic. Microscopic study classes them as examples of parenchymatic nephritis, becoming transposed to a condition of interstitial nephritis.

The rectum and bladder were not diseased. The intestines were normal, with the exception of the vermiform appendix, which was gangrenous from its terminal extremity almost to its origin in the cæcum. There was, however, no loss of structure or perforation. It contained no concretions or fecal matter.

I was unable to exactly determine why this gangrenous condition existed, had the child survived a few days longer, perforation would undoubtly have occurred. The condition of the appendix may possibly be explained by recalling the anatomy of the part.

The mesentery of the appendix is derived from the inferior layer of the mesentery of the ileum, and is too short, ending usually at about the centre of the appendix, or, at most, at the junction of its middle and terminal third.

There is another structure, which Treve aptly states, is the remains of the true mesentery of the appendix. It takes its origin from that part of the ileum most remote from its mesenteric attachment, and is united with the mesentery of the appendix. This peritoneal fold does not carry any blood supply to the appendix, which as a rule,

Fig. 3.

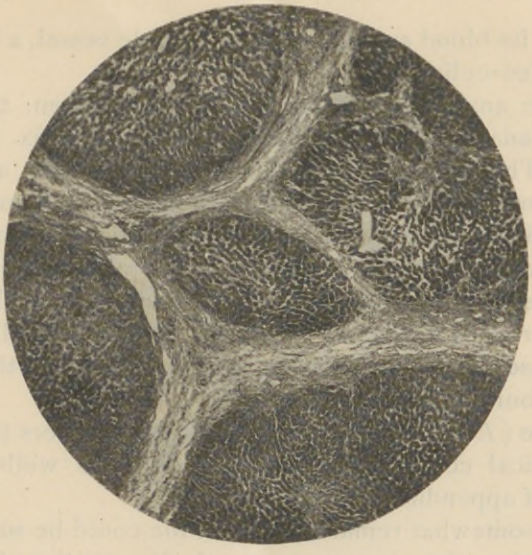

Fig. 4

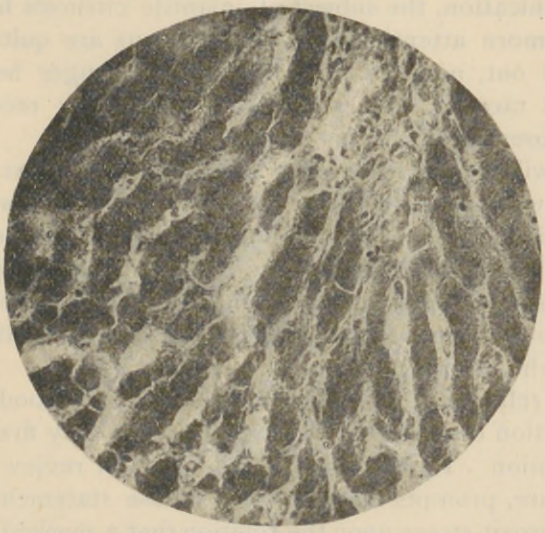

FIGURE 3. SHOWS LOBULE SURROUNDED BY THE ENORMOUSLY INCREASED INTERLOBULAR CONNECTIVE TISSUE. (x 35 DIAMETERS.)

FIGURE 4. SHOWS THE SAME LOBULE MORE HIGHLY MAGNIFIED. (x75 DIAMETERS.)

derives its blood supply from but a single vessel, a branch of the ileo-colic.

These anatomical facts, would appear then, to offer an explanation of the condition of the appendix in this case. The enormous quantity of peritoneal fluid, and the distention of the cæcum, head of the colon, or ileum, with gas, and the interruption of intestinal peristalsis from fluid pressure, would cause dragging on the already too short mesentery of the appendix. In this way, we would have a material interference with the blood supply, and per consequence, an exciting causative factor in the condition found at the post-mortem.

White (*Therap. Gaz.*, June 15, 1894), considers that the anatomical condition completely explains well-known types of appendicular disease.

It is somewhat remarkable, that life could be sustained with such an important organ as the liver in the advanced state of disease shown by the specimen. The organ was practically converted into a mass of fibrous tissue.

Since the appearance of Howard's paper and my first communication, the subject of infantile cirrhosis has received more attention, and its problems are quite well worked out, nor can the affection any longer be considered rare, as writers are now constantly recording their observations in this disease.

We will briefly review the progress since 1890. The recent writings have shown us, that the natural history of cirrhosis of the liver in childhood does not differ from the same disease in the adult. Its cause, clinical symptoms, diagnosis and prognosis are identical. Alcoholism seems to have the same relation to the disease in early life as in the cirrhosis of adolesence or advanced age.

The relation of the eruptive fevers of childhood to the production of cirrhosis, was dwelt upon in my first communication. Further study, and a careful review of the literature, prompts me to reiterate these statements, and to lay great stress upon the relation that a marked attack of any of the eruptive fevers has upon the production of the disease. This relationship was clear in the case which

presents the specimen under consideration, and other writers have recorded similar observations, particularly is this the conclusion of the Thesis of Porembski, "Contribution a l'etude des Cirrhosis Hepatiques ches les Enfants." Paris, 1891.

In Stack's* twenty cases at St. Bartholomew's Hosptial, and the Hospital for Sick Children, London, occurring in children under twelve years of age, averaging five years, there was no evidence that alcohol, syphilis or rickets was either directly or indirectly the cause; but, on the other hand, seven of the cases had suffered from scarlatina. In five, the symptoms of cirrhosis were decided to have dated definitely from the attack of scarlet fever. Six cases had measles, but the relation between the causative effect of this exanthema and cirrhosis, was not as clear as in those who had scarlatina. Acute nephritis was coincident in some instances. Jollye reports two fatal cases (*Brit. Med. J.*, Apr. 23, 1892, *Am. J. Med. Sci.*, Oct., 1892,) brother and sister, aged ten and eleven, who had both previously suffered from measles. Ormerod (St. Bartholomew's Hospital Reports, Vol. xxvi, 1890,) reports a remarkable case of cirrhosis of the liver in a boy, with obscure and fatal nervous symptoms. The child was aged ten. The post-mortem showed lesions in the brain and advanced hepatic cirrhosis. Vertical sections through the brain showed at the level of the optic commisure a smal patch of softening, involving the outer layer of the left lenticular nucleus. The section, one and one half inches further forward, (pediculo-frontal), and the section behind, (through anterior border of pons the parietal section), no longer showed this softened patch. Some tiny patches of softening occurred in the upper part of the pons. No disease of the cortex cerebri or of cortex cerebelli. Ormerod refers to other cases in the literature, which presented the same peculiar and fatal nervous symptoms with post-mortem evidence of advanced hepatic cirrhosis and slight lesions in the nervous system.

The writer thinks that the Germans have presented

* *Am. J. Med. Sci.*, Vol. civ., No. iv., p. 501.

Fig. 5.

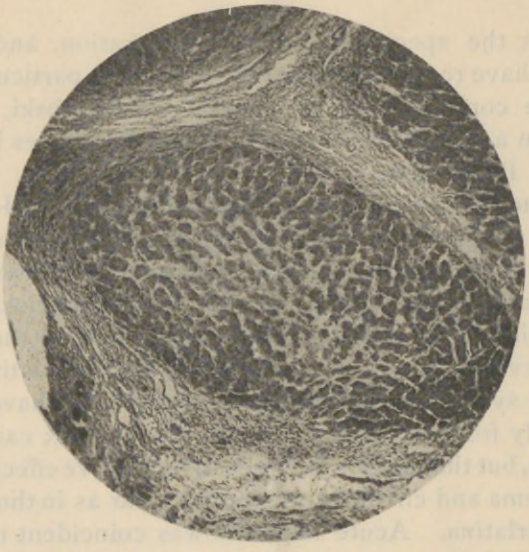

Fig. 6.

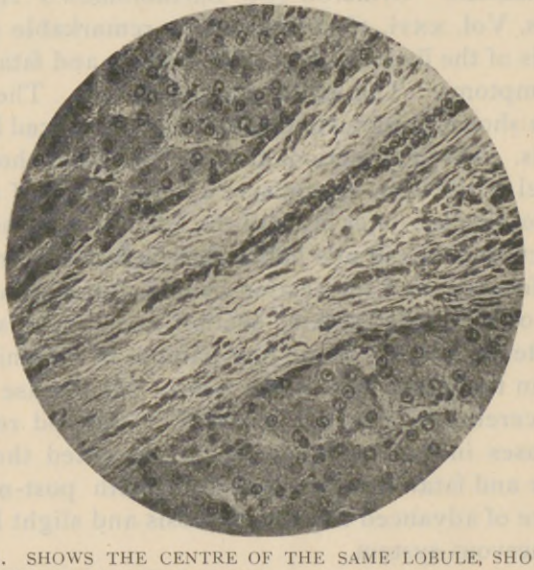

FIGURE 5. SHOWS THE CENTRE OF THE SAME LOBULE, SHOWING INCREASED INTRALOBULAR TISSUE.

FIGURE 6. SHOWS THE EDGE OF THE SAME LOBULE, SHOWING THE DENSE CHARACTER OF THE INCREASED TISSUE.

evidences, which make it probable that the softening in the neighborhood of the lenticular nuclei had some essential connection with the disease. Gowers' case of cirrhosis of the liver presented symptoms intermediate between those of chorea and tetany, but no lesion was found in the nervous system after death. Tordeus, *Jour. de Med. de Chir. et de pha. Bruxelles*, Nov. 29, 189– (have not seen the original) reports a case in a boy of nine years, in which no predisposing or exciting cause could be found.

Some late writers have considered hepatic cirrhosis as infectious in its origin. Neumann, *Deuts. Med. Zeit.*, Berlin, Mar. 23, '93 (Cohen) reports a case of congenital hepatic cirrhosis, probably dependent upon syphilis. Death occurred at four months of age. Hausemann, (*Wien. Klin. Wochenschrift*, May 8, '93,) referred to by the same reviewer, also reports a congenital case. D'Espine (*Sajou's Annual*, c. 45, Vol. 1, 1894,) records a case of cirrhosis of the liver in a boy who came under observation at the age of six years, and was observed for three years. Jaundice or albuminuria never occurred. Ascites was present for two years; repeated right-sided pleural effusions. Necropsy showed extensive fibroid changes and adhesions of peritoneum and viscera, perihepatitis, perisplenitis, enlargement of spleen. The liver exhibited evidences of an intense, interstitial, periportal hepatitis. Bouchard, in commenting on the case, combatted the idea that infantile cirrhosis was due to syphilis.

Jollye (*ibid.*) remarks that the habit of the inordinate consumption of vinegar by the two children under his care, had in his opinion, much to do with the production of the cirrhosis. The vinegar lessening gastric secretion, interfered with digestion, and allowed the formation of albuminoses and allied bodies, and by absorption into the portal system, set up a hyperplasia of the connective tissue in the liver.

Stack (*ibid.*) formulates the following conclusions:

"That alcohol, syphilis, tuberculosis and malaria, account for 50 per cent. of the cases, the other most fre-

quent cause being probably the exanthemata and errors in diet.

"That acute interstitial hepatitis is frequently found microscopically after the infectious fevers, especially after measles and scarlet fever ; but the part played by the disease, alcohol, and diet, respectively, in those cases which afterward become examples of cirrhosis, is an open question, as is also the reason why some livers are affected with hypertrophic and others with the simple form.

"That some cases are part of a general disease, due to some poison gaining admission to the general circulation, and especially attacking the liver owing to the slow circulation in the hepatic capillaries, just as no doubt, acute yellow atrophy is a general disease, the chief pathological change found *post-mortem*, having caused it to be classified amongst the diseases of the liver."

Brown's interesting case (*Arch. Pediat.*, Vol. X., No. 1., 1893, p. 48,) in a boy aged nine years and nine months, with careful post-mortem records, seems to have been due entirely to dietetic excesses.

Ghosh (*Indian Med. Gaz.*, January, 1891; Am. J. Med. Sc., May, 1891,) in reporting cases of cirrhosis in children under two and a half years of age, that almost invariably terminated fatally in from three to twelve months, proposes as the only etiological element a faulty diet.

Osler states that in the cirrhosis of early life, excluding the alcoholic and syphilitic cases, the acute infectious diseases are probably the important antecedents.

The more recent papers of the year just closed are as follows :

Blagoveschenski, A. N. "Atrophic Cirrhosis of the Liver in a Child of Ten Years." *Trudi Obst. dietsk, Vrach.*, Mosk., 1894, 11,129.

Mirinescu. "Cirrhose Hypertrophique avec ictére Chronique Chez un Garcon de 14 Ans." *Rev. Mens. d. Mal. de l'Enf.* Paris, 1894, xii., 560–567.

Sainsbury, H. "Alcoholic Cirrhosis of Liver in a

Child ; Ascites ; Meningitis ; Death ; Necropsy." *Lancet*, London, 1894, 1, 147.

Penrose, F. "Cirrhosed Liver from a Youth Aged Sixteen." Tr. Path. Soc., Lond., 1892–3, xliv., '93.

Clarke, J. M. "Remarks on Cirrhosis of the Liver, with Especial Reference to its Occurrence in Children, and to the Mode of Death in Cirrhosis with Jaundice." *Brit. M. J.*, Lond., 1894, 1, 1407–1412.