PERICARDITIC PSEUDO-CIRRHOSIS OF THE LIVER, OR PICK'S DISEASE.

BY

A. WILSON GILL, M.D. Ed., M.R.C.P.,
SENIOR HONORARY ASSISTANT PHYSICIAN, NORTH STAFFORDSHIRE
ROYAL INFIRMARY;

AND

T. CRAIG, M.B., CH.B. GLAS.,
HOUSE-PHYSICIAN TO THE HOSPITAL.

THE association in young people of enlarged liver with recurring ascites and chronic adherent pericardium is not a very common condition. The earliest reported case was probably the one reported by Van Deen in 1846, but a more complete description was given in 1896 by Pick, and the symptom-complex is often described as Pick's disease, or Pick's pericarditic pseudo-cirrhosis of the liver. Most modern textbooks of medicine do no more than merely mention this disease, but many monographs have been written on it, and most of the references in this paper are abstracted from a lengthy monograph by Kelly¹ printed in the *American Journal of the Medical Sciences*.

The disease presents many problems yet unsolved, notably the true cause of the condition and the relationship it bears to two other diseases—namely, Concato's disease on the one hand, and "Zuckergussleber," observed by Budd in 1852, and graphically named "sugar-iced liver" by Curschmann in 1884, on the other.

In Pick's disease chronic adherent pericardium occurs in association with chronic venous congestion of the liver, and while few adhesions are found in the peritoneum, the surface of the liver, though often opaque, does not show the dense layer of fibrous tissue common in sugar-iced liver. Rolleston,² in fact, declares that there is a very definite microscopic distinction in these two conditions. The fibrous tissue increase in Pick's disease is deposited under the capsule; whereas in Curschmann's disease the fibrous overgrowth is outside the capsule. The same author also states that "if peritoneal adhesions are advanced, the condition ceases to be Pick's pseudo-cirrhosis of pericarditic origin," and belongs to that of chronic universal perihepatitis associated with adherent pericardium. Kelly also has some difficulty in classifying the three conditions which clinically and pathologically seem to merge into one another, and considers that they are best grouped under the wider term of "polyserositis" or "polyorrhomenitis."

The important factor in all is the occurrence of chronic inflammatory changes in various serous surfaces, associated with a great tendency to ascites; this demands frequent tapping, although the intervals between each paracentesis may be far apart. In a case described by Rumpf the ascites was tapped 301 times, and in Osler's³ case of a child 121 times. This factor alone is of importance in distinguishing the disease from the true multilobular cirrhosis, which rarely requires tapping more than a few occasions before death ends the disease. In other respects the etiology is different from cirrhosis chiefly in respect of the age incidence—polyserositis tending to attack young people—and in the absence of the usual irritants—for example, alcohol. Pick recognized this difference when he called his disease "pseudo-cirrhosis," but the name is not a bad one, as when the cases are seen first the diagnosis of cirrhosis is often made, especially as the adherent pericardium is sometimes difficult to diagnose during life. When the disease occurs in young people an alternative diagnosis of tuberculous peritonitis is often made, and in this connexion Rolleston and others⁴ consider that the tubercle bacillus may play a part in the causation.

The pathological findings in polyserositis are fairly constant and have been reported many times. In fully developed cases changes are found in all the serous surfaces, although the changes in the pericardium and round the liver are the most obvious and most constant. Dense adhesions, often calcified, are found in the pericardium, valvular disease is often present, and the heart enlarged. Adhesions occur in both pleural cavities, the right side

being usually more advanced than the left. Enlarged spleen with perisplenitis and chronic peritonitis occur. The liver is usually enlarged, and shows the usual nutmeg character of chronic venous congestion. Microscopically, the liver shows irregular areas of fibrosis, but generally the amount of fibrosis is scanty and may be absent in considerable areas (Rolleston).

Most observers have been puzzled, in view of the pathological findings, to explain the recurring ascites, and many theories have been advanced. The frequent occurrence of dense adhesions in the right pleural cavity was believed by Weinberg, in 1887, to cause angulation and distortion of the inferior vena cava and hepatic veins, and Eisenmenger supported this theory in 1900. Neither Kelly nor Rolleston favours this view. Harris (1895) considered that the ascites was the result of a peculiar form of chronic peritonitis, while others believe that the heart condition is responsible. Against this latter view is the usual absence of oedema of the legs, although in some cases a transient oedema occurs at the beginning of the illness. Kelly sums up the three opinions thus: (1) That the ascites is due to changes in the liver, the result of long-standing congestion. (2) That the ascites is due primarily to the chronic peritonitis. (3) That the ascites is due to the combined action of these factors.

The actual exciting cause of the condition is still obscure. In many cases there is no history of antecedent illness. In others there is some indication of an acute onset attacking one serous membrane—for example, the disease may follow a definite attack of pleurisy or an attack of pericarditis. It has occasionally followed certain acute and infectious illnesses, notably rheumatism, syphilis, typhoid fever, and malaria. Both Kelly and Rolleston lay great stress on antecedent rheumatism. Others believe that the tubercle bacillus is an important factor even in the absence of definite evidence of tuberculosis, and some authors actually state that all cases of chronic adhesive pericarditis are tuberculous in nature. Certainly it is a fact that some of these cases ultimately die of tuberculosis. Picchini (1901) distinguished four groups of cases.

1. Cases where the origin of the disease can be determined—that is, the first serous membrane affected.
2. Cases where the origin may be determined, but in which the first examination of the patient reveals some affection already of several serous membranes, although they manifest no clinical signs.
3. Cases where the history reveals that from the beginning several serous membranes have been affected.
4. Cases where it cannot be determined whether the disease began with one or more serous membranes.

Mention has already been made of the prevalence of the disease in youth, and the following figures from Kelly's paper demonstrate this fully. Of 39 cases, 21 occurred in males, 18 in females; 3 were under 10 years of age, 13 were between 11 and 20 years, 3 were between 21 and 30 years, 5 were between 31 and 40 years, 11 were between 41 and 50 years, and 4 were between 51 and 54 years.

The following case exhibits most of the clinical features common to such cases—namely, the early age incidence, the absence of antecedent disease, the gradual onset, the occurrence of ascites and enlarged liver, in association with evidence of cardiac disease, presumably adherent pericardium.

A boy, aged 14½ years, was admitted to the North Staffordshire Royal Infirmary on June 26th, 1928, because of ascites and throbbing of the neck.

History.—In June, 1927, he began to notice an increasing size of the abdomen, which caused him to have some slight shortness of breath on exertion, so that he no longer could challenge boys to run races or play football with vigour. Closer questioning elicited the fact that pulsation in the neck on the right side had been noticed by his parents for the previous two years, but no importance was placed by them on this observation as they looked upon him as a perfectly fit boy, who had played football for the school for two seasons. He left school at Christmas, 1927, beginning work as an apprentice engineer in January, 1928, and working steadily until June 5th. On this date various apprentices were referred to the factory surgeon for examination as to fitness, when this boy was rejected and advised to secure medical advice. For the purpose of this examination he cycled a distance of three miles, and on arrival was obviously short of breath. He then consulted Dr. A. Gilchrist of Burslem, who diagnosed "Concato's disease" and referred him to the Royal Infirmary.

Previous Illnesses.—He was born at full time, was bottle-fed, and was not looked upon as a delicate child. When 2 months old he had a slight attack of bronchitis or broncho-pneumonia lasting two weeks, from which he made a full recovery and had no complications. At the age of 7 years he developed measles, and again there were no ill effects. He has suffered from occasional colds in the head and slight rheumatic pains in the wrists in winter, but there is no history of tonsillitis, chorea, or acute rheumatism, and no record of pleurisy. At the age of 12 he slipped off a 4-foot wall to the ground and landed with his knee pressed against his chest. He was at once short of breath, and had pains across the chest which lasted three hours, after which he felt quite well and returned to school the next day. Since measles at the age of 7 he has not missed school attendance.

The family history is not important, the parents being alive and healthy, three brothers alive all healthy, and no deaths in the family have occurred. There is no tuberculous history on either side.

Symptoms on Admission.—Fullness of the abdomen and throbbing of the neck were the obvious symptoms. Occasional pain across the chest and in the left shoulder and slight indigestion were further admitted. No cough was complained of, and no record could be obtained of swelling of the legs.

Condition on Examination.—His height was 5 ft. 1½ in., his weight 7 st. 10 lb. He did not look a robust boy, the complexion being somewhat sallow, but there was no cyanosis and no obvious shortness of breath. Undue venous pulsation was noticed in the neck, especially on the right side. No clubbing of the finger-nails was present and no oedema of the legs. The abdomen was distended by free fluid, the circumference at the umbilicus being 32 inches. The free edge of the liver could be felt three inches below the costal margin, the liver itself feeling smooth but firm. Scanty pubic hair was present, but there was no hair in the axillae. A few small glands were present in the axillae. The teeth were good, the tonsils not enlarged, tongue slightly coated. On July 8th paracentesis was performed, 96 ounces of a clear straw-coloured fluid being withdrawn. Culture was negative, and very few lymphocytes were found on staining. Injection into a guinea-pig had no ill effects on the latter. After withdrawal the lower pole of the spleen was just palpable. No enlarged glands or tuberculous masses could be felt in the abdomen. The chest was hyper-resonant in the upper lobes. The lungs expanded freely. The breath sounds throughout were harsh vesicular in type. No adventitious sounds were heard except a fine friction over the splenic area. The chest was x-rayed on June 26th by Dr. J. Bromley of Stoke-on-Trent, who reported as follows:

"Both diaphragms are raised, doubtless due to the amount of free fluid in the abdomen. There is no marked increase in the extent of the hilum shadow and no impaired air entry into the chest." The urine was free from albumin and sugar, and the urea concentration test was normal. Blood urea did not exceed 13.5 mg. per 100 c.cm. The Wassermann reaction of the blood was negative. A fractional test meal revealed no abnormal features. The levulose tolerance test increased the amount of sugar in the blood from 0.12 to 0.22 per cent., and no sugar was passed in the urine. The blood count on June 26th showed red blood cells 3,700,000, white blood cells 11,500, haemoglobin 75 per cent. Differential count: Polymorphs 80 per cent., lymphocytes 18 per cent., eosinophils 1 per cent. No abnormal cells were found.

Condition of the Heart.—A distinct fullness was observed over the precordium. The apex pulsation was scarcely visible, and there was no indrawing of the interspaces. No thrill was felt on palpation, and the positions of the heart borders and of the apex beat itself were difficult to define by percussion, owing to the hyper-resonant note of the lungs. Careful percussion, however, seemed to locate the apex beat in the fifth interspace about half an inch outside the nipple line. There was a definite epigastric pulsation visible. Auscultation revealed a curious high-pitched musical squeak, which was audible at one time and absent at another. The character of this bruit was very peculiar. On certain occasions it was heard all over the precordium and over the left scapula; at other times it was absent behind. Sometimes it was more marked at rest, sometimes only after exercise. It was at times loudest during deep expiration, and occasionally it would disappear altogether for long spells, during which intervals no valvular disease was apparent. The timing of this sound also varied. Usually it coincided with the closure of the aortic valves, at other times it occurred during ventricular systole. It seemed to have its origin outside the heart itself, and probably depended upon some adhesion between the pericardium and neighbouring structures. The systolic blood pressure was consistently low, the average being 112 mm. of mercury. The pulse rate averaged 90 per minute, and the exercise tolerance test was normal. The electrocardiogram taken both before and after paracentesis showed the constant inversion of T-wave in leads II and III. No particular ventricular preponderance was present.

Progress.—After paracentesis on July 8th the boy's health seemed to suffer and a progressive slight loss of weight occurred for a week or two afterwards. With iodide of iron internally and iodoform inunction to the abdomen his health began to improve and he gained weight—some of this increase being due no doubt to a slight return of the ascites. The chest was again x-rayed by Dr. J. Bromley on August 10th, his report being: "There appeared to be some enlargement of the ventricular shadow to the right, while in the left hilum a distinct bulge is seen, suggesting an enlarged clump of glands. Both diaphragms moved well at this examination." A further blood count was performed on August 22nd, which showed the improvement which had occurred—namely: red blood cells 5,500,000, white blood cells 16,500, haemoglobin 85 per cent. Differential count: Polymorphs 78 per cent., transi-

tional and mononuclears 4 per cent., lymphocytes 16 per cent., eosinophils 1 per cent., basophils 1 per cent. No abnormal red cells were found.

He was discharged on September 1st in good condition, and when re-examined on November 4th, 1928, he looked plump and well, but ascites had largely accumulated again and a very slight oedema of the legs was found. His weight was 8 st. 3 lb., and there was no apparent shortness of breath or distress on exertion. Further paracentesis will shortly be necessary.

REFERENCES.

¹ Aloysius O. J. Kelly: On Multiple Serositis, etc., *Amer. Journ. Med. Sci.*, cxv, 1903, p. 116. ² H. D. Rolleston: *Diseases of the Liver, Gall Bladder, and Bile Ducts*, 1912, p. 96. ³ Osler and McCrae: *Modern Medicine*, vol. iii, 1925, p. 786. ⁴ G. W. Norris and H. R. M. Landis: *Diseases of the Chest*, third edition, 1924, p. 724.

INOPERABLE CARCINOMA OF THE BREAST TREATED WITH RADIUM.

BY

ROY WARD, M.B., B.S.,

ASSISTANT MEDICAL SUPERINTENDENT, THE RADIUM INSTITUTE, LONDON.

In considering the radium treatment of carcinoma of the breast it must be remembered that both external applicators and needles are employed. The apparatus must be screened in such fashion as to cut off the beta rays, which are harmful to the tissues, and permit the gamma radiation to pass; 2 mm. of lead is recommended for external plaques when prolonged exposures are contemplated. A compound known as "Columbia wax" is most useful when treating a large surface area. For slow-growing tumours needles with walls of 0.5 mm. of platinum are recommended; these vary in active length from 1 to 5 cm., and contain 0.5 mg. of radium element to each centimetre. More recently tiny needles known as "seeds" have been found to be useful in some cases. The most convenient seed is one containing 2 millicuries of radon and screened through 0.5 mm. of platinum or gold. In cases of encephaloid character, where the growth is extremely rapid and very vascular, more powerful needles, screened through 1 mm. of silver, are often of great value.

REVIEW OF CASES.

An attempt has been made to ascertain the fate of the 633 patients suffering from breast carcinoma who have been treated at the Radium Institute during the last ten years. A study of these cases has not only made it possible to form an opinion as to the value of radium therapy in hopeless cases, but from the information obtained certain principles can be formulated for the treatment of patients in whom the disease is less advanced. In the series under review, which does not include patients who received prophylactic treatment, it was found that in every case the disease had extended to tissues outside the breast itself.

TABLE I.—Showing Type of Cases and Site of Metastases.

	Total No. of Cases.	Primary Inoperable Cases.	Recur- rent Cases	Axillary Glands.	Supra- clavicular Glands	Chest Wall.	Ulceration.	Oedema of Arm.
1918	82	16	66	43	44	25	4	11
1919	88	21	67	39	45	21	4	10
1920	81	29	52	31	33	33	10	19
1921	69	16	53	31	29	32	11	12
1922	41	6	35	19	23	15	6	5
1923	44	10	34	23	11	11	12	5
1924	39	12	27	23	16	13	3	10
1925	66	15	51	36	34	25	11	10
1926	58	21	37	24	27	21	8	11
1927	65	18	47	31	34	25	14	12
	633	164	469	300	290	223	83	105