RECURRENT HAEMATEMESIS IN "BANTI'S DISEASE"

BY

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The physician who discusses the splenic anaemia syndrome finds himself in an uneasy and apologetic position. This is of course because the pathologist is able to make the statement that primary Banti's disease does not exist. But at the bedside the use of the term must be conceded to cover those instances of splenomegalic anaemia for which none of the many known causes can be found. This position is supported by Whipple's (1937) record of a group of thirty-one patients with "Banti's disease." In no fewer than fifteen of these it was not possible to find the causal factor. So that, as Castle and Minot (1936) put it, the problem posed by Banti still remains unanswered. There are three usual explanations employed "to empty the Banti pigeon-hole"—cirrhosis of the liver, obstruction of the splenic vein, and hepatico-lienal fibrosis. It is generally held that sections of these spleens reveal changes resulting from long-standing congestion. Whipple uses the term "portal hypertension," and notes that work in his clinic shows that a very high pressure occurs in the splenic vein; and the recollection of the tense bulging splenic veins seen at operation supports this. So that the present attack on Banti's disease is on the vascular and mechanical fronts. But before opening the question of haematemesis I would like to make a few more general points.

In the Liverpool Royal Infirmary records over the last ten years it has only been possible to find twenty cases where there was enough positive and negative evidence to fill the classical criteria. The main facts are embodied in Table I.

TABLE I.—Twenty Cases of "Splenic Anaemia" at the
Liverpool Royal Infirmary (12 men, 8 women;
average age, 32.7 years)

Average Blood Picture:

Haemoglobin, 48 per cent.; red cells, 3,500,000 per c.mm.; white cells, 4,200 per c.mm.; lymphocytic trend.

Platelet Counts:

Thrombocythaemic, 2 cases; normal limits, 2 cases; thrombopenic, 15 cases; no counts, 1 case. Average count in thrombopenic cases, 100,000 per c.mm.

Fractional Test Meals (12 Cases):

Low normal, 4; almost achlorhydria, 1 case; achlorhydria, 7 cases.

Haematemesis occurred in 9 and splenectomy was performed in 7 cases.

It would probably be agreed that the blood picture shown there is the one commonly found. Yet it must be emphasized that such haematological evidence is a shaky diagnostic basis. The red cells and the haemoglobin can be restored to normal by administering massive doses of iron (Davidson, 1934). This must be related to the frequent achlorhydria seen in these cases and those in the literature; though it is not known whether the achlorhydria is histamine-fast. The link between the splenomegaly and the absence of acid is still missing, as is the reason why both the giving of iron and the removal of the spleen correct the anaemia. The leucopenia almost always present in "splenic anaemia" occurs in other splenomegalies and is not diagnostically specific. The platelet count is seen to be low in most cases, but it must be remembered that recent haematemesis will cause an increase to normal

figures: the existence of occasional normal and high counts is again an unreliable diagnostic criterion. The dangers of splenectomy in other than thrombopenic cases have been conclusively shown by Rosenthal (1925) and confirmed by Howel Evans (1928). But with these views Whipple (1937) has expressed disagreement. Because of the relationship of two of the patients cited in this paper an attempt was made to find recorded instances of "familial splenic anaemia," but without success. It was possible to examine six members of this family, but there was no other splenomegaly or anaemia. The only abnormality in the examination and investigation of these people was a persistent thrombopenia in the father of Case 3. Negative though this evidence is, the occurrence in first cousins is probably more than a coincidence.

Splenectomy and Haematemesis

Single or repeated haematemesis is of course a well-known feature of the splenic anaemia syndrome, but it is not so well known that splenectomy does not always remove the liability to this type of bleeding. It had been thought that once the patient had survived the immediate post-operative dangers, the later risk of thrombosis, and the possibility of hepatic cirrhosis he was out of the wood. But most clinicians have had an experience similar to that which aroused my own interest—the rapid and fatal exsanguination of a man apparently doing very well two years after splenectomy. Pemberton (1931) puts this into figures when he records that this symptom recurred post-operatively in 50 per cent. of a group of ninety-eight patients with "splenic anaemia" who bled before the operation. Death was a result of this haemorrhage in twenty-nine patients, against nine from thrombosis and four from hepatic cirrhosis in the larger group of 182 cases of "splenic anaemia." Similarly Dr. Howel Evans, who has followed a smaller series, has seen recurrence after operation in about half the patients and death in several. But it is important to note that only those who bled before removal of the spleen bled afterwards. That these further bleedings are due to growth of the hepatic obstruction is an obvious solution. If we study the figures supplied by Whipple (1937) we will see that much of the recurrent haematemesis is found in the group with hepatic cirrhosis. But I would suggest that though this explanation is obvious it is not the only factor at work. If it were we should have to issue automatically a bad prognosis in these recurrences, and so overlook the patients in whom, as I hope to show, the problem is a simple mechanical one.

If a mouthful of rather stiff barium paste is allowed to slip down the oesophagus of patients who have vomited blood either because of cirrhosis of the liver or "splenic anaemia," the distended oesophageal varices can be outlined radiographically. Kirklin and Moersch (1931) in reporting this confirmed the varices by oesophagoscopy. These veins may extend throughout the lower two-thirds of the gullet; they are large, tense, and immediately submucous. A liability to bleed is obvious, though there may occasionally be another danger spot in distended gastric veins. Kegaries (1934) has shown that the venous bed at the lower end of the oesophagus has three connexions: directly from the portal system via the coronary vein along the lesser curvature of the stomach; indirectly from the splenic pedicle by the vasa brevia; and to the superior vena cava via the azygos veins. In hepatic cirrhosis, then, there will be dilatation of all the veins in the portal system. And the liver will be "by-passed" by a distended coronary vein to oesophageal varices. In "splenic anaemia" without cirrhosis of the liver the venous congestion in the splenic bed will cause the vasa brevia to open up and varices will be formed by the second mechanism. The coronary vein has been tied with good results—as regards haematemesis—when dilated and without result when of normal size (Rowntree, Walters, and McIndoe, 1929). But it

cannot yet be stated (Walters, personal communication) whether the size of the coronary vein can determine the site of venous obstruction, though it is likely that the size of this vein at operation will be a guide as to whether there is enough venous block in the liver to produce oesophageal piles. There is no need to discuss here the possibility of superior vena cava block causing varices; but it must be clear that once the veins in the lower end of the oesophagus become distended they are columns of blood in direct connexion with the superior vena cava and unprotected by valves.

Operation for Relief of "Splenic Anaemia"

With these details of the vascular mechanisms in mind it is possible to examine the various operative methods of relieving "splenic anaemia." An example of the effect of splenectomy on a patient who had shown previous haematemesis is seen in Case 1. The effect of ligation of the coronary vein has already been dealt with. Warner (1929-30) has reported the result of tying the splenic veins in a boy with "Banti's disease" showing a large liver and ascites. The rationale was to lessen the blood flow through the spleen, and there was a marked immediate improvement. Splenectomy was subsequently necessary, and the patient has been well for the last five years (Warner, personal communication). Ligation of the splenic pedicle has been suggested by McNee (1931), by Graham Bryce (1932), and by Howel Evans (1936). Watson (1935) has reported a successful instance of this operation very similar to Case 2. The patient was a woman, the usual criteria of "splenic anaemia" were present, and she had reached the stage of ascites. Because of adhesions splenectomy was not feasible and the whole pedicle was tied. The spleen became very tender and there was a post-operative thrombosis, but ultimately recovery was complete. Three years later there were no symptoms. The blood picture being slightly subnormal in all respects—as in Case 2—Case 3 is presented as another example of the results of pedicle ligation.

Case 1

A furnace-man, aged 32, was admitted under the care of Dr. A. Douglas Bigland in October, 1935. There was no familial splenomegaly and no residence abroad, and the only previous illness was typhoid fever at the age of 15. There had been a haematemesis six years before, another three years before, and one just before admission. A sallow pallor had slowly appeared, and there had been some symptoms of anaemia. The spleen was found to be one fingerbreadth below the costal margin with no hepatomegaly or ascites. No free hydrochloric acid was produced by a gruel test meal. The barium meal showed no gastro-intestinal abnormality. Dr. Howel Evans reported the blood picture to be: red cells, 3,400,000 per c.mm.; haemoglobin, 35 per cent.; leucocytes, 1,125 per c.mm.; platelets, 122,000; and halometer reading, 7.17 microns. The bleeding and clotting times were unchanged, the fragility normal, and the serum Wassermann negative. By means of large doses of iron for two months the haemoglobin was raised to 80 per cent. and the red cells to 5,000,000. Since several platelet counts showed a consistent reading in the neighbourhood of 100,000 splenectomy was decided on and was carried out by Mr. Hugh Reid. Section of the spleen showed the "diffuse fibrosis usually associated with Banti's disease." In the post-operative period the platelet count did not rise above 700,000 per c.mm. and no thromboses occurred. The patient returned to work soon afterwards and remained well even without iron.

For follow-up purposes he was seen ten months later; he felt very well, was working, and there had been no recurrence of bleeding. The haemoglobin was 100 per cent., the red cells 5,600,000 per c.mm., the leucocytes 12,800 per c.mm., with slight lymphocytosis, reticulocytes 1.5 per cent., and halometer

reading again 7.17 microns. The platelet counts were 165,000 and 186,000 per c.mm. on successive days. On giving a gruel test meal there was again no free acid secreted, but a moderate amount appeared after histamine. Search for oesophageal varices, which was not specially made prior to the operation, showed these to be present and distended sufficiently to be easily recognized by a barium swallow.

Case 2

This patient, a boy of 14 who has for many years been under the care of Mr. J. T. Morrison, to whom I am indebted for permission to publish the case, is an only son and first cousin to Case 3. There was no other instance of splenomegaly or anaemia in the family, and those of his relatives I was able to examine were all normal. From 1929 to 1931 he was in hospital most of the time with the sequelae of an appendix abscess and empyema. There was then no splenomegaly, and two blood counts showed a secondary anaemia and leucocytosis. From 1932 to 1935 there were four haematemeses, and he was in hospital several times. Clinically there was anaemia, some stunting of growth, and a spleen enlarged to an inch below the costal margin. A typical blood count was: red cells, 3,300,000 per c.mm.; haemoglobin, 45 per cent.; leucocytes, 2,500 per c.mm.; and platelets, 137,500 per c.mm. The serum Wassermann was negative, as were those of his parents; the bleeding and clotting times and the blood fragility were within the normal ranges. Operation was refused until 1935, by which time ascites had appeared. When the abdomen was opened the spleen was found to be so adherent that removal would have been impossible. Mr. Morrison therefore tied the splenic pedicle vessels. Recovery from operation was rapid, and the boy gained his proper weight and height. Within a few months he was able to play games for the first time for several years.

I examined him two years later. He was then a well-grown boy, but the olive tint of the face persisted. The rounded splenic edge could be felt on inspiration. The gruel test meal revealed small amounts of free acid. The blood picture (no iron had been given since the operation) showed: red cells, 4,624,000 per c.mm.; haemoglobin, 95 per cent.; leucocytes, 5,680 per c.mm.; reticulocytes, 1.5 per cent.; and platelets, 190,000 per c.mm. The fragility was unaltered. The radiologist was able to show well-filled oesophageal varices.

Recently, after four days of sore throat and pain on swallowing, another large haematemesis occurred, following which the spleen shrank so as to be almost impalpable. The liver was not enlarged, and there was no ascites or other evidence of portal cirrhosis. The oesophageal veins were again seen radiographically. Here once more the dangers to which the varices are exposed—perhaps this time there was an oesophagitis—were emphasized.

Case 3

A married woman, aged 24, was admitted to the Liverpool Royal Infirmary in September, 1936, under the care of Dr. Bigland. She had never been abroad and had had no previous illness of note. Despite this she had always been pale and tired. During the last five years there had been flatulent dyspepsia, and food seemed to stick at the level of the lower end of the sternum, and for the last year she had suffered from dyspnoea and substernal effort distress, possibly due to the anaemia. Just before admission she had vomited about two pints of blood. The facies was olive-tinted, the heart showed a basal haemic bruit, and a firm-notched spleen reached to the umbilicus. Dr. Howel Evans reported the blood picture as: red cells, 4,260,000 per c.mm.; haemoglobin, 50 per cent.; leucocytes, 2,250 per c.mm.; platelets, 136,000 per c.mm.; with pallor but no microcytosis of the red cells. The serum Wassermann was negative, and the clotting time, bleeding time, and fragility were within the usual limits. The stomach tube could not be swallowed, but the barium meal showed no gastric abnormality. Extensive oesophageal varices were present. For the next three months the patient was

treated with large doses of iron and weekly injections of liver extract. The details of the various blood counts are shown in Table II (personal observations). It was clear that the case was thrombopenic in type and suitable for splenectomy. The patient, however, preferred the simpler operation of ligation of the splenic pedicle, such as had been so successful with her cousin. Since it seemed to us that this procedure could do no harm ligation was deliberately undertaken. Mr. Hugh Reid opened the abdomen and found the liver macroscopically

in the oesophagus were unaltered. Intensive iron therapy was prescribed, and slow improvement is taking place.

In addition to these three cases it was possible to examine radiologically two other patients who had been subjected to splenectomy for classical "Banti's disease." In a young man who had had haematemesis both before and since operation there were extensive varices throughout the lower two-thirds of the oesophagus. In a middle-aged woman, in whom there had been no bleeding but in whom hepatic cirrhosis had been found at operation, no varices were to be seen.

The results of pedicle ligation will be of interest to those who advocate the operation. The general effect in Case 2 can be described as excellent, since the lad has grown to his proper height and weight and is able to follow a normal life. Locally the spleen has shrunk remarkably and cirrhosis of the liver has not appeared. Yet his life is hazarded by a problem in vascular mechanics. In Case 3, where the ligation was more deliberately carried out, the result was not so good, but further improvement may be seen. Though the red cells and haemoglobin are kept at healthy levels with the use of iron, there are still many general symptoms. The spleen has been reduced, but it is even now too big to be regarded as inactive. The varices, and therefore the risk of further haematemesis, seem unaltered. There was a good deal of post-operative pain and reaction, as would be expected with such a drastic interference with the blood supply. To counterbalance this there is probably a lower immediate risk, and certainly from the low platelet rise much less danger of thrombosis. But on the whole the patient would have been better off with splenectomy, and may even yet need it. From this small experience one might suggest that ligation of the pedicle may be useful in two situations: where the platelet level is normal or high and splenectomy is likely to be followed by a thrombocythaemia of over 1,000,000 per c.mm.; and at those laparotomies where the adhesions are too thick for safe splenectomy.

It becomes clear, then, that the post-operative study of this type of patient shows that whether the dilated veins at the cardia are formed either from portal obstruction or from a more local obstruction they persist after splenectomy and pedicle ligation. The fact that half the individuals with previous haematemesis do not bleed again is probably due to lessening or stoppage of the blood flow upwards through the *vas brevia*. That the other half do bleed again is due to the direct communication between the varices and the superior vena cava. Portal obstruction is therefore not essential to maintain the distension. No doubt the bigger the veins are before operation the more likelihood there is of subsequent haemorrhage. It is a fair analogy to point out that cure of varicose veins in the leg is not attained by distal ligation. The proximal ligation or removal of oesophageal varices is a matter of great technical difficulty and beyond my province. Walters and Kegaries (1933) have suggested the submucous injection of sclerosing solutions. But one may conclude that in those instances where haematemesis follows splenectomy, and haemorrhage, not cirrhosis of the liver, is likely to be the cause of death, the purely mechanical problem of dealing with the varices will have to be faced.

Summary

It has been shown that, because of the persistence of oesophageal varices after operation in cases of "Banti's disease," neither splenectomy nor splenic pedicle ligation will prevent recurrence of haematemesis.

TABLE II.—*Serial Blood Counts in Case 3*

Date	Red Cells (millions)	Leucocytes (per c.mm.)	Platelets (per c.mm.)
9/9/36	Haematemesis—two pints		
10/9/36	4.24	3,200	170,000
11/9/36	4.00	5,900	222,000
14/9/36	4.16	3,200	91,000
16/9/36	3.68	4,000	81,000
18/9/36	4.08	2,800	53,000
21/9/36	4.16	4,000	65,000
23/9/36	4.00	3,200	63,000
25/9/36	4.26	2,250	136,000
28/9/36	4.32	2,800	96,000
1/10/36	3.20	5,600	106,000
	Out-patient—liver and iron		
6/12/36	5.40	5,660	168,000
8/12/36	5.20	—	161,000
9/12/36	5.20	4,750	98,000
16/12/36	5.12	6,520	94,000
17/12/36	Operation		
18/12/36	5.30	16,000	200,000
19/12/36	5.36	16,000	200,000
20/12/36	5.44	14,600	367,000
21/12/36	5.60	9,600	523,000
22/12/36	5.40	—	745,000
23/12/36	4.80	11,500	300,000
24/12/36	5.04	—	504,000
25/12/36	5.20	6,200	196,000
29/12/36	4.80	—	216,000
30/12/36	5.20	—	400,000
31/12/36	4.64	—	—
1/1/37	4.16	12,800	249,000
2/1/37	4.20	11,200	550,000
3/1/37	4.24	—	424,000
5/1/37	4.96	—	291,000
7/1/37	4.70	—	450,000
9/1/37	Discharge. Convalescence with iron		
19/4/37	5.16 (Hb, 85%)	4,800	129,000
1/5/37	5.90 (Hb, 95%)	8,000	147,000

normal, the coronary vein not dilated, but the *vasa brevia* larger than usual. The splenic artery was tied in two places and the three splenic veins, each as large as one's thumb, were occluded. The post-operative period was stormy, and a good deal of pain arose from the tender spleen. Convalescence was slow, but four months after the operation the patient was able to get about, though pale and suffering from lassitude. The spleen, while considerably shrunken, was still two inches below the costal margin. Though the red cells were over 5,000,000 per c.mm. the haemoglobin was only 85 per cent., and leucopenia and thrombocytopenia were still present. The radiologist was able to show that the varices

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THE ASSOCIATION OF TRAUMA WITH PROGRESSIVE MUSCULAR ATROPHY

BY

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We know little indeed about the aetiology of progressive muscular atrophy, but its association with trauma in a small percentage of cases has been described by eminent Continental neurologists during the past decade. Thus Alajouanine, Gopcevitich, and Meillaud (1930) gave an account of an interesting case of post-traumatic muscular atrophy, with an illuminating discussion on the subject. These authors divide such atrophies into different groups.

The Muscular Atrophy Groups

In the first group the muscular atrophy is localized to the region of the muscles in the neighbourhood of the injured joint. They state that to this group belong some of the amyotrophies of the periscapular region which may follow injuries to the shoulder-joint, and suggest that such amyotrophy is of reflex origin, as is indicated by the electrical changes and by the fact that after progressing for several months cases remain stationary or even retrogress. They define this type as "localized post-traumatic reflex amyotrophy."

In the second group the amyotrophy ensues rapidly upon the injury, and it develops on a paralytic background. Its distribution is generally radicular, and it is accompanied by sensory changes of a dissociated character which suggest that this type may follow minor traumatic fissures of the spinal cord, with resulting frustrated haematomyelia. In such cases the wasting, at first progressive, becomes stationary after a time. They describe it as spinal amyotrophy secondary to frustrated haematomyelia.

In a third group of cases the wasting develops more slowly. It has not been preceded by any notable paralytic phenomena and is not associated with sensory troubles or reflex abnormalities. The atrophy in such cases may be localized or diffuse and non-systematized. The electrical reactions are not myelopathic in type, but conform closely to the changes found in the myopathies. This group progresses for a time, but may later display a tendency to regression.

Descriptive Cases

Alajouanine and his colleagues describe the case of a man, aged 34, who was seen at the Salpêtrière for a diffuse muscular atrophy which had slowly progressed over a period of twelve years. The condition had begun two months after an aeroplane accident which had rendered him unconscious, and in addition to contusions had resulted in a split fracture of the head of the radius.

Another case of post-traumatic muscular atrophy is described by Horst Geyer (1934). This patient sustained a fractured tibia and fibula of the right leg in 1905. Four years later wasting was observed in the right thigh. Clubbing of the right foot was also present as a result of acute anterior poliomyelitis which had occurred in infancy. When examined by Horst Geyer in 1932, in addition to the typical claw hands, wasting and fibrillary tremors were widespread. The ankle and knee reflexes were practically absent, and there was no evidence of any pyramidal tract lesion. The small muscles of the hands and feet showed no response to galvanism or faradism.

The next case described by Horst Geyer is that of a male, aged 52, who in 1921 was struck in the small of the back by a branch of a tree. As a result of this he was unable to work for several weeks, but subsequently recovered. Eight years later he fractured his left leg in a motor-cycle accident. When examined at the clinic on May 9, 1930, the left leg showed marked generalized wasting. No abnormalities were observed in the rest of the body. When seen again three years later there were definite signs of progressive muscular atrophy, wasting and fibrillary tremors being present in all the limbs, though they were more marked on the left side than on the right.

Predisposing Factors

Geyer remarks that these two cases of his have in common the onset of progressive muscular atrophy some years after the original injury. In discussing the frequency and pathogenesis of the condition, he says that of the twenty-one cases which appeared at the clinic between the years 1919 and 1933 only one could be shown clearly to be due to trauma. He quotes Kure, who stated in 1931 that the innervation of muscles is fourfold—cerebral, spinal, sympathetic, and parasympathetic—and disturbance of any of these can lead to muscular atrophy. He points out, further, that many eminent neurologists are of the opinion that the main feature is a congenital predisposition, and that trauma has merely "pulled the trigger" in such cases. This theory, he says, is held by Strümpell, Probst, Oppenheim, and others. Marburg asserts that this is merely a possible theory, and that there is no anatomical evidence in support of it. Other authors maintain that one has to consider the question of an acquired disposition. Rose divides the predisposing factors into five groups: one is the simple type previously mentioned; the other four are acquired—namely, toxic; infective; those occurring after cerebrospinal diseases, such as anterior poliomyelitis; and, finally, the traumatic group, such as those associated with fractures. Other authors also stress the association with anterior poliomyelitis in some cases. Thiern considers that there occurs in the traumatic cases a traumatic neuritis which spreads up towards the spinal cord, and eventually by association with the anterior horn cells produces progressive muscular atrophy. Another theory propounded is that, acting on the congenital or acquired predisposition, there occurs in the traumatic cases a shaking or