SPLENIC ANAEMIA OF YOUNG CHILDREN TREATED BY SPLENECTOMY.

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We wish to draw attention to the possibility of treating cases of splenic anaemia in young children by the removal of the spleen, and to record three cases treated by splenectomy with successful results.

Splenic anaemia of young children, or, as it is sometimes called, von Jaksch's anaemia, is a disease which occurs during the first three years of life and is characterized by marked anaemia, great enlargement of the spleen, and much general debility. The disease is essentially chronic and continues for many months, or even years. The prognosis in advanced cases is unfavourable, though some do undoubtedly improve and recover after prolonged treatment. Many are carried off by an intercurrent disease, such as bronchopneumonia, to which they are very liable.

Splenic anaemia of young children has no relation to the splenic anaemia of adults, and it is probably a disease *sui generis*, though the exact cause is not yet known. Some observers are of opinion that it is an extreme form of rickets with anaemia. Most cases of splenic anaemia of young children do show some signs of rickets, but this is not always well marked. The disease may occur in twins; of this we have seen two instances.

The symptoms are insidious, accompanied by indigestion, and it is impossible to say exactly when the disease actually begins, but the child gradually becomes pale and wasted and the enlarged spleen is then detected. The pallor is marked and the colour is often pale lemon. The red blood corpuscles are much diminished in number in a severe case and may fall to just over 1,000,000 per c.mm.; they vary in size and shape and there are many nucleated red corpuscles; the haemoglobin is much reduced. There is a moderate increase in the white cells, with a preponderance of lymphocytes and a few myelocytes. In a severe case the spleen reaches down to the pelvis and stretches across the middle line; it is hard and the notch is well felt. The liver is increased in size. The lymphatic glands are not enlarged. There is generally pyrexia, mild but prolonged.

TREATMENT.

The treatment is on general lines, with proper feeding combined with arsenic and iron. X-ray treatment has been of little value in our hands and has seldom influenced the course of the disease or the size of the spleen. Owing to the uncertain results obtained with medical treatment in advanced cases, and the fact that these cases often do not improve but rather tend to grow worse, we decided to try the effect of removal of the spleen, and have performed this operation on three occasions during the past year. Splenectomy was only undertaken after medical treatment had failed to give relief and after careful consideration.

It seems that the spleen in these cases is a diseased organ, and that the child will be better without it if it can be removed with a reasonable degree of safety. Splenectomy is now recognized as a definite surgical procedure, and the operation has in recent years largely increased its scope. Until fairly recently surgical removal of the spleen was limited to cases of injury, torsion, or where it was the seat of a new growth; now, however, it is performed in certain cases of anaemia, cirrhosis, and acholuric jaundice, and appears to give excellent results in suitable cases.

It is well established that the spleen is not essential for the carrying on of life, and little harm seems to come to the

individual after its removal. In those cases of splenomegaly the etiology of which is at present unknown, splenectomy appears to be a procedure of distinct clinical value. Before undertaking it every effort must be made by the careful preparation of the patient to reduce the risk of the operation as far as possible. Owing to the extreme degree of anaemia from which these patients may be suffering, their condition can be much improved and the blood count correspondingly raised by preliminary transfusions of blood. In addition, when the spleen is considerably enlarged, the re-transfusion of the blood it contains, after the spleen has been removed, is a procedure worthy of consideration. The blood so obtained is unaltered in character, and there appears to be little risk in this simple method of auto-infusion. Further, the application of a massive dose of x rays over the spleen shortly before operation, as advised by Moynihan,¹ may lead to shrinkage of the organ and render the operation more simple to carry out. In one of our cases the reduction in the size of the spleen under the influence of x rays was marked.

In performing splenectomy a left-sided paramedian incision, with outward displacement of the rectus muscle, was used, and this gave ready access to the organ. Adhesions were troublesome in two of the cases, but with careful clamping the spleen was removed without difficulty and with very little haemorrhage. The absence of post-operative shock in such young children was very noticeable and was largely due to the careful preliminary treatment of the patients. The following are the notes of the cases.

CASE I.

A boy, aged 1 year and 11 months, was sent to the hospital from an infant welfare clinic on account of anaemia. The child had been breast-fed for the normal time, but since the age of 1 year he had not been well. For the five months previous to coming to hospital there had been difficulty with his feeding, some vomiting, and a marked loss of weight. There had been no cough, no abdominal pain, and the bowels were regular. The spleen was very large, reaching well down into the pelvis and across the middle line of the abdomen. It was hard and the notch was easily palpated. The child was of a pale lemon colour and the blood count was:

Red blood corpuscles	...	...	...	1,360,000
White blood corpuscles	...	...	...	31,200
Polymorphonuclears...	...	...	...	26 per cent.
Lymphocytes	...	...	...	71
Haemoglobin	...	...	...	30

A fair number of nucleated red corpuscles and a few myelocytes were seen. The blood platelets were increased. The Wassermann reaction was negative and there were no haemorrhages.

He was admitted to hospital on April 22nd, 1925, where he was kept for a month. He was properly dieted, had a course of arsenic and iron, and also x-ray treatment to the spleen. He improved only slightly and was sent home, where the same treatment was continued. He became rather worse, and was readmitted to hospital on June 17th, 1925. The spleen was causing trouble because of its great bulk, and the child could hardly bend forward. He seemed to us as likely not to recover, and it was decided to remove the spleen. The blood count at this time was practically the same as when admitted to hospital on the first occasion. He was slightly rickety.

On July 2nd, as the father proved to be a suitable donor, the child was given a transfusion of 40 c.cm. of blood into the external jugular vein. Blood examination a week later showed that the red blood corpuscles numbered 1,420,000 per cubic millimetre. Two subsequent transfusions from the father of 60 c.cm. each were given on July 11th and July 17th, and the blood count showed that the red cells had risen to two and a half millions. On July 19th he received a massive dose of x rays over the enlarged spleen.

On July 20th splenectomy was carried out. The spleen, which appeared to occupy about half the abdomen, was easily delivered and adhesions caused little trouble. The pedicle was divided between clamps and the organ handed to an assistant. By squeezing the organ 40 c.cm. of blood were obtained from the splenic vessels, and this was run into citrate solution and returned into the external jugular vein of the child. There was no free fluid in the abdominal cavity, and the liver, which was slightly enlarged, appeared smooth and normal in colour and consistence.

The spleen, after removal, weighed 540 grams (18 oz.), and it was then devoid of blood. The length was 15 cm. and the width 10 cm. It had preserved its normal form; it was fairly firm and

its surface was smooth. The colour was a uniform chocolate or reddish-brown. The capsule was not thickened. Microscopically there was much fibrosis.

Post-operative History.

The child stood the operation remarkably well and the next day he was happy and taking his food as usual. There was no rise of temperature and practically no post-operative shock. Four days after operation the red blood cells numbered 2,400,000. Recovery was uneventful, and he left hospital some weeks later much improved in general health. Blood examination on August 8th showed:

Red blood cells	...	...	...	4,300,000
White blood cells	...	...	...	15,000
Polymorphs	...	...	...	48 per cent.
Lymphocytes	...	...	...	48 "
Large mononuclears	...	...	...	2 "
Eosinophiles	...	...	...	2 "
Haemoglobin	...	...	...	72 "

There were no nucleated red cells.

The child has been kept under close observation since then, and he has improved very much. He is nearly a normal child for his age; he can walk and his colour is good. The blood count on December 23rd, 1925, was as follows:

Red blood cells	...	...	...	4,800,000
White blood cells	...	...	...	6,062
Polymorphonuclears	...	...	...	48 per cent.
Lymphocytes (small)	...	...	...	50 "
" (large)	...	...	...	2 "
Haemoglobin	...	...	...	85 "

CASE II.

A boy, aged 1 year and 5 months, was admitted to hospital early in August, 1925, with the typical symptoms and signs of splenic anaemia. He had been ill with indefinite symptoms for several months. There was no evidence of rickets and the Wassermann reaction was negative. The spleen was very large and extended down to the pelvis and across the middle line at the level of the umbilicus. The accompanying photograph was taken prior to operation to indicate the size of the spleen. The blood count on admission was as follows:

Red blood cells	...	...	...	2,320,000
White blood cells	...	...	...	16,400
Haemoglobin	...	...	...	50 per cent.

The red cells showed some anisocytosis, and there was a large increase in the number of cells larger than the average normal. There was a moderate degree of metachromasia, and fairly numerous punctate granular forms were present. There were many nucleated forms of several

types, the predominant type being normoblasts. A differential count of the leucocytes gave the following results:

Polymorphonuclear cells	...	...	...	52 per cent.
Lymphocytes	...	...	...	45 "
Large mononuclear cells	...	...	...	2 "
Eosinophil cells	...	...	...	1 "

A fair number of myelocytes were seen.

The liver was moderately enlarged and there were no haemorrhages. As he had not improved after two months' medical treatment and x rays it was decided to remove the spleen. The same procedure as in the first case was carried out prior to operation, and the spleen was removed with little difficulty on September 30th. The spleen, after removal, had retained its normal shape, was fairly firm, dark reddish-brown in colour, but rather more of a chocolate hue than the normal organ. The capsule was not much thickened; there was no infarct or haemorrhage. The cut surface was uniformly dark reddish-brown. Its weight was 215 grams (devoid of all blood) (7 oz.), length 13 cm., width 8.15 cm., thickness 3.5 cm.

The child made a rapid convalescence and showed signs of improvement within a few days after the splenectomy. His subsequent progress has been most satisfactory in every way. A blood count on December 23rd showed:

Red blood corpuscles	...	...	...	5,260,000
White blood corpuscles	...	...	...	4,890
Haemoglobin	...	...	...	85 per cent.

CASE III.

A boy, aged 2 years and 3 months, had been treated by his own doctor for anaemia and general weakness for several months. He

was admitted to hospital in August, 1925, suffering from marked anaemia and an enlarged spleen and a blood count as follows:

Red blood corpuscles	...	...	...	2,300,000
White blood corpuscles	...	...	...	13,000
Haemoglobin	...	...	...	50 per cent.

The red cells showed some anisocytosis and there was a slight excess of cells of larger size than the average normal. There was a moderate degree of metachromasia. Some finely granular punctate basophilic cells were present. Nucleated forms of two or three different types were fairly numerous. A differential count of the leucocytes gave the following result:

Polymorphonuclear cells	...	...	...	64 per cent.
Lymphocytes	...	...	...	32 "
Large mononuclear cells	...	...	...	3 "
Eosinophiles	...	...	...	1 "

A few myelocytes and mast cells were present. The Wassermann reaction was negative. He showed no apparent signs of rickets.

He remained in hospital for six weeks, where he had medical and x-ray treatment, and he was then sent home slightly improved. After returning home he gradually became worse again, and was readmitted to hospital at the end of November. The spleen at this time reached to the level of the umbilicus and the blood count was practically identical with that in August. The liver was moderately enlarged and there were no haemorrhages.

In view of the apparent success following removal of the spleen in the previous two cases, it was decided to perform splenectomy, which was done on December 11th. He received blood transfusions and a massive dose of x rays prior to operation, as in the other cases. He made an uneventful recovery, and he is now, as far as can be judged, a normal child. A blood count on January 6th, 1926, showed:

Red blood corpuscles	...	...	...	4,737,000
White blood corpuscles	...	...	...	5,675
Haemoglobin	...	...	...	80 per cent.
Colour index	...	...	...	0.98

PROGNOSIS.

In a series of twelve cases treated by one of us on medical lines, seven died, giving a mortality of 58 per cent. Of von Jaksch's² original three cases, one ended fatally, whilst Aschenheim and Benjamin's³ record four fatal cases out of five. Thursfield states⁴ that 65 to 70 per cent. recover, giving a mortality of 30 to 35 per cent. Rolleston⁵ quotes Beretervide and Bianchi, who give a gloomy prognosis, and state that in ten years they have not seen a case recover after medical treatment. The prognosis under medical treatment is so unsatisfactory, and the outlook so serious, that the effect of surgical removal of the spleen on the course of the disease certainly seems justifiable. This operation has been tried in America⁶ with encouraging results in a few cases.

The immediate improvement in all three cases here recorded leads us to hope that this form of treatment may have a definite place in dealing with this disease. The ultimate results in these cases will be carefully watched and recorded at a subsequent date.

CONCLUSIONS.

Splenectomy appears to have a definite place in the treatment of advanced cases of splenic anaemia in early childhood, after medical treatment has been tried for some time and found to be unsatisfactory.

Splenectomy brings about an apparent rapid cure, though its ultimate value must be awaited for a further period of time and until many more cases have been so treated.

The risks attached to the operation, after a careful preparation of the patient, do not appear to be great.

We wish to report three cases of splenic anaemia in young children treated by removal of the spleen after medical and x-ray treatment had been found to be unsatisfactory. All the cases have recovered and appear to be greatly benefited by the operation.

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REFERENCES

- 1 Moynihan: BRITISH MEDICAL JOURNAL, July 11th, 1925, p. 47.
- 2 Von Jaksch: *Prag. med. Woch.*, 1890, xv.
- 3 Aschenheim and Benjamin: *Deut. Arch. f. klin. Med.*, 1912, C.V. 470.
- 4 Thursfield: *Diseases of Children* (Garrod, Batten, and Thursfield).
- 5 Rolleston: BRITISH MEDICAL JOURNAL, December 12th, 1925, p. 1101.
- 6 Stillman: *Amer. Journ. Med. Sci.*, vol. cliii, 1917.