HIRSCHPRUNG'S Disease or Megaloclon is a condition characterized by symptoms and physical signs which follow a fairly constant pattern, but whose aetiology is still not wholly proven, and whose treatment is far from satisfactory.

The disease, usually seen in infants and children, may appear in adults, presenting a similar clinical picture, though usually of a milder degree. Boys are more often affected than girls. Briefly the typical history is that of a male child suffering from obstinate constipation, with periodic attacks of diarrhoea. Vomiting may occur, growth is stunted, and the child is of sallow complexion. The abdomen is grossly distended. Diagnosis is confirmed by a Barium enema, or a Barium meal with a follow-through.

Pathology

The dilatation and hypertrophy are confined usually to sigmoid and descending colouir; there is no organic obstruction distal to the affected gut, the gut below the recto-sigmoid junction being normal in calibre. The hypertrophy results from an increase in the circular muscle fibres, this at first being the chief change; later ulceration of the mucosa from stagnant faeces leads to infection and secondary fibrosis. It is obvious that any treatment likely to lead to normal bowel calibre must be performed before secondary changes take place, i.e., in the very young.

Aetiology

Most authorities regard the cause of megaloclon as being
due to neuromuscular incoordination, an imbalance between the sympathetic and parasympathetic taking place, leading to overaction of the sympathetic.

That this theory is probably correct is suggested by the results of various sympathectomy procedures and by the occasional association with other conditions, such as megaloureter, whose etiology is regarded as having a similar basis.

**Treatment**

In the earliest stages this is essentially medical, liquid paraffin and regular daily enemata, distressing both to mother and child, being its chief features. Also various drugs, both parasympathetic stimulants and paralysants, have had their sponsors.

**Surgical Treatment**

Excision of the affected bowel was widely practised prior to any form of operation on the sympathetic system. This procedure is still performed but must inevitably carry a very high mortality.

**Sympathectomy**

Various procedures have been put forward, each with its own peculiar disadvantages. Left lumbar ramisectomy, lumbar 11 to lumbar v, was first advocated, and this was subsequently widened into lumbar ganglionectomy, with resulting loss of sympathetic control to the vessels of the lower limb.

Presacral neurectomy with prolongation upwards to include the origin of the inferior mesenteric artery from the aorta, and excision of the inferior mesenteric ganglion was the next to be tried. This operation is prone to cause sterility, by failure of ejaculation, a serious complication, especially as males predominate in this condition.

**Spinal Analgesia** as a form of treatment was first suggested by Stabins, Morton and Scott in 1935. Previously this had been used as a preliminary test as to the probable efficiency of a sympathectomy. They found that the mere giving of a spinal anaesthetic would restore the normal peristaltic mechanism of the bowel. Hawksley found that out of twelve cases so treated
eleven showed marked improvement. This avoided the dangers of an open operation in these children, but there still existed the dangers associated with a high spinal anaesthetic. In order to be certain that all splanchnic fibres are included, a high spinal block to the level of D₄ has to be performed.

In view of the reported favourable results following spinal analgesia, the authors decided to try the effects of splanchnic block on these patients, as this seemed to have some advantages over spinal block.

The fall of blood-pressure following splanchnic block is much less than that following a high spinal anaesthetic, and other complications which may follow splanchnic block are fewer and less serious than those which may follow spinal block.

The technique which was used in this series, Kappis method of posterior splanchnic block, may on occasions present some difficulties, especially in children, where the landmarks cannot always be identified with ease, but it is frequently less difficult to perform than the lumbar puncture necessary for the spinal anaesthetic.

In this series Anathaine or Amethocaine Hydrochloride with Adrenalin was the drug used; the strength and quantity of the solution varied with the age and weight of the patient.

**Case 1. Adult male aged 46 years**

Splanchnic block was performed under local analgesia. 80 cc. of 1/4000 Anathaine Solution were injected each side. There was no fall of blood-pressure following the injection and the patient's general condition remained satisfactory. He was treated with daily enemata and purgatives. On the 14th day after the splanchnic block, his bowels acted without an enema; after this his symptoms were entirely relieved and his bowels acted regularly each day. Two years later he was readmitted to hospital for a partial gastrectomy. The operation was successfully performed under Regional Analgesia; at operation his colon still showed hypertrophy and dilatation. He remained symptom-free during his stay in hospital, and also after his discharge.
When he was last seen, 3½ years after the splanchnic block had been performed, he was in good health and showed no symptoms of his megalocolon.

Case 2. Female aged 17 years

Splanchnic block was performed under local analgesia. 80 cc. of 1/4000 Anathaine Solution were injected into each side. She experienced no fall of blood-pressure or other symptoms following the block. Daily enemata were given, and on the 11th day after splanchnic block her bowels acted without an enema. She was discharged symptom-free, and has remained so for 3 years.

Case 3. Female aged 11 years

Splanchnic block was performed under light general anaesthesia. 50 cc. of 1/1000 Anathaine Solution were injected each side. Her blood-pressure dropped from 120/100 to 110/90 within half an hour of the completion of the splanchnic block, but rose rapidly after that. She showed no other symptoms, and her condition gave no cause for anxiety. After daily enemata and purgatives her bowels acted on the 12th day without an enema. She was discharged symptom-free, and has remained so for 3 years.

Case 4. Male child aged 4 years

Splanchnic block was done under a general anaesthetic. 11 cc. of 1/2000 Anathaine Solution were injected each side. He showed no fall of blood-pressure or other symptoms, and was given daily bowel wash-outs. On 11th day, as his bowels had not yet acted without an enema, the splanchnic block was repeated under a general anaesthetic. On this occasion 10 cc. of 1/1000 Anathaine Solution were injected. Recovery was uneventful; four days later his bowels acted without an enema and his abdominal distension was considerably relieved; he was discharged from hospital one week later, his constipation entirely relieved. He has remained symptom-free for 18 months.
CASE 5. Male infant

Splanchnic block first performed under a general anaesthetic at age of 8 months; 1/6000 Anathaine Solution was used, but the result was unsatisfactory. He was a weak, undersized child, and was retained in hospital for many months, receiving treatment for various ailments unassociated with megalocolon. At the age of 2 years the splanchnic block was repeated under a general anaesthetic; 10 cc. of 1/2000 Anathaine Solution were injected each side. He showed no ill effects following the splanchnic block, and was given daily bowel wash-outs. The block, however, proved a failure and his constipation was not relieved, so 21 days later the block was again repeated under a general anaesthetic. 10 cc. of 1/1000 Anathaine Solution were injected each side on this occasion; and he showed no immediate ill effects following the injection. Four days later, in spite of daily bowel wash-outs, he was still constipated, and started vomiting—he also showed marked abdominal distension. After several large, hard faecal masses had been removed from his colon his condition improved, the abdominal distension became less, and he stopped vomiting. From this time his bowels acted without an enema at irregular intervals, but though his condition had improved the results of the splanchnic block did not appear satisfactory. At this stage he developed measles and was transferred to an isolation hospital. In view of the apparent failure of the block it is intended to readmit him at a future date for further treatment.

The treatment of four of the five patients in this series has proved satisfactory. These patients have now been symptom-free from 18 months to 3 ½ years. The failure of the splanchnic block in Case 5 was probably due in part to the difficulties in technique, in this undersized child, also probably to the fact that the correct strength and quantity of Anathaine Solution necessary to give a satisfactory result has not yet been injected in this instance.

In spite of the daily enemata and purgatives given to these patients, no result occurred until between the 11th and 14th
day after the splanchnic block, and as yet there seems to be no obvious reason for this.

The authors consider that as splanchnic block, as a method of treatment for Hirschsprung’s Disease, has proved satisfactory in these patients, and as it appears to have fewer dangers and complications than most other forms of treatment used for this condition, it is worth while trying it before using more drastic forms of treatment.

**Summary**

The aetiology, pathology and treatment of Hirschsprung's Disease are discussed, and the splanchnic block is suggested as a method of treatment for this condition. A summary of five cases treated by this method is given and some advantages discussed.

**References**