"IDIOPATHIC" DILATATION OF THE COLON.

"HIRSCHSPRUNG'S DISEASE."

Thesis for the degree of M.D.

by

D. P. D. WILKIE,
M.B., Ch.B., F.R.C.S.E.

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"IDIOPATHIC" DILATATION OF THE COLON.
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The name Idiopathic dilatation of the Colon originally given by Gee to the condition which I propose to discuss in the following pages is an unsatisfactory one in that while aiming at being descriptive it is so vague as to permit of somewhat free interpretation. In studying the literature on this subject it early becomes evident that the term "Idiopathic" conveys to different minds different ideas, so that from the very outset one has to work within ill-defined limits, and this laxity of definition asserts itself and presents difficulties throughout the entire study of this subject.

The German name "Hirschsprung's disease" applied to the same morbid condition has now become even less precise in its meaning, than has the English name. The Italian synonym "Megacolon Congenito" is much more definite, but in the opinion of most authorities it implies more than is justified by our present knowledge of the disease it refers to. Probably the name Hirschsprung's disease is as good as any, for while it is always now associated with a morbid condition giving rise to a characteristic clinical picture and and showing very pronounced pathological changes, it/
it is frankly non-committal as to etiology in regard to which certainty is as yet impossible.

**Definition.** By Hirschspring's disease we understand a dilatation, with hypertrophy of the walls, of the whole or some part of the large intestine, no organic obstruction of the bowel distal to the dilated portion being present.

This dilatation and hypertrophy is usually (not always) most pronounced in the pelvic colon, and is frequently limited to that portion of the great gut. The degree of the dilatation and hypertrophy is generally very great before the case is diagnosed, and the dilated coil of bowel frequently appears to fill almost the whole abdomen.

The condition is usually met with in young children, many of the cases reported being under one year of age, but cases have presented themselves for treatment at almost all ages and one of the cases which I had the opportunity to study was a man over 60 years of age.

Pronounced constipation dating from birth or shortly after birth characterizes the condition, and abdominal distension and enlargement are marked features. The majority of the recognized cases have died /
died in early childhood from intestinal toxaemia or from an attack of acute enteritis. It is not uncommon however to find the disease in older children.

The fact that quite a number of cases have been met with in adults past the prime of life, shows probably that milder degrees of this condition may cause so few symptoms during childhood and early adult life as to escape notice, and only when the degenerative changes of old age begin to set in does the condition assert itself.

At present it is almost impossible to form an accurate opinion as to the frequency with which this disease occurs, as it is only within recent years that its existence has been recognized. It is a significant fact, however, that in each successive year a larger number of cases is reported than in any previous year, and the total number in the literature now exceeds 100.

The disease however is much commoner than this figure would suggest as a great number of cases never find their way into the generally available medical literature.

All statistics go to show that the disease is much commoner in boys than in girls. Duval collected/
collected forty-five cases and thirty-six of these were boys. Later statistics entirely support this proportion.

Historical.

In 1846 Dr. Carlo Favalli described a case which must be taken as one of Hirschsprung's disease, in a man, fifty years of age, where post mortem an enormous degree of hypertrophy and dilatation of the pelvic colon and to a less degree of the rest of the large intestine was found.

In 1851 Little and Calloway reported a case of enormous dilatation and hypertrophy of the colon in an imbecile thirty-four years of age. The enlargement involved chiefly the descending and pelvic portions of the colon which filled more than half of the abdomen.

In 1868 A. Jacobi reported a case where in an infant there was complete intestinal obstruction from birth. Under the impression that the case was one of atresia of the rectum operation was undertaken. The child died immediately after the operation. At the sectio it was found that the obstruction was due to a long multiply flexed and kinked pelvic colon and descending colon above which the meconium had collected. (The reason for including this as a case/
case of Hirschsprung's disease will be explained later.)

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In 1872 Peacock described a case of fatal constipation from excessive dilatation of the colon. The patient twenty-eight years of age had been constipated from birth and had suffered much from flatulence. He enjoyed relatively good health till within six weeks of his death when obstructive symptoms set in and the abdomen became greatly distended. Post mortem - the entire colon was found greatly dilated.

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In 1884 Gee published his communication on "Idiopathic Dilatation of the Large Intestine". He gave as an illustrative case that of a boy of four and a half years who had suffered from constipation since the age of three months and whose abdomen continued to swell from the time he was a year old.

It was not till 1888 however that Hirschsprung published his first two cases of this condition and gave for the first time a clear clinical picture of the disease.

In his original paper he held that there could be/
be no doubt about the congenital origin of the condition which he took to be an abnormally great development of the large intestine and not a dilatation and hypertrophy due to obstruction of any kind.

In 1890 Hirschsprung published two further cases and in 1901 four more cases.

Since 1890 cases have been reported from all quarters and quite a considerable literature has accumulated on the subject. Though Hirschsprung's original description of the clinical and pathological conditions has been little altered, his view, that the condition is always truly congenital has been stoutly contested by many observers.

Numerous theories have been propounded with regard to the etiology of this affection; the one which has found especial favour is that which may be called the "Anatomical Theory," as it holds that an exaggeration of the relatively long and lax pelvic colon, found normally in the infant, is the predisposing cause of this condition.

It will be advisable therefore at the outset to consider carefully the anatomy of this part of the large intestine, especially in the child.
ANATOMY OF THE PELVIC COLON.

In the Adult.

The pelvic colon is a large coil of intestine which begins at the inner border of the left psoas muscle where it is continuous with the iliac colon and ends at the level of the third sacral vertebra by passing into the rectum. Between these two points it has a well developed mesentery and forms a large and variously shaped coil which usually lies in the cavity of the pelvis. Its most common arrangement in the pelvis is the following.

Beginning at the inner margin of the left psoas it first plunges over the brim into the pelvis and crosses this cavity from left to right; it next bends backwards and then returns along the posterior wall of the pelvis towards the middle line, where it turns down and passes into the rectum.\(^6\)

The Pelvic meso-colon is a fan-shaped fold short at each extremity and long in its middle portion. Its root is attached along an inverted V shaped line one limit of which runs upwards and inwards as high as the bifurcation of the common iliac artery (sometimes higher); here it bends at a fairly acute angle and the second limb descends over the front of the sacrum.
to the middle of its third piece where the mesentery ceases and the pelvic colon passes into the rectum. Fossa Intersigmoidea.

When the pelvic colon with its mesentery is raised upwards, at the apex of the V shaped attachment one usually finds a small pocket or recess. This may be so small as hardly to admit the tip of the little finger, but it has been found much larger, in several cases extending up as far as the pancreas. It is due to the imperfect blending of the mesentery of the descending meso-colon with the pareital peritoneum of the posterior abdominal wall.

Fig. 1.

Intersigmoid Fossa

Distended Pelvic Colon Thrown upwards. Pointer indicated into Intersigmoid Fossa which in this case extended upwards almost to the Pancreas.
Plica Inguino-colica.

This is a fold of peritoneum rising in the region of the crural canal and running upwards along the inner border of the Psoas to lose itself on the under surface of the pelvic meso-colon. (Engel7) This fold only comes into existence when the pelvic colon and its mesentery are raised upwards and put on the stretch, as when that part of the bowel is distended. Many anatomists do not recognize or describe it, on the ground that it is an artificially produced fold; the constancy with which it is present, however, especially in children, justifies its description. I found it present, sometimes well marked, sometimes only just recognizable, in everyone of fifty cases of newly born infants and young children which I examined.

Ligamentum Mesenterio-Mesocolicum.

This peritoneal fold was first described by Gruber8 as follows. It is a fold of peritoneum which stretches from the upper surface of the lower part of the meso-sigmoid upwards and to the right obliquely across the vertebral column presenting a sharp sickle like edge. It generally arises from a point two thirds to three quarters way along the meso-sigmoid/
meso-sigmoid (i.e. nearer its rectal end). If the ileum be pulled over towards the right side, the fold can be seen running obliquely across the body of the fifth lumbar vertebra to the mesentery of the ileum and it may stand out as much as one to two inches high. Usually the band is four to five inches long but sometimes it is not more than two inches. When the small intestines are thrown upwards and to the right the free edge looks upwards and forwards. With the intestines in situ it looks forwards and downwards. In a few cases the band was found to rise from the middle of the meso-sigmoid or even nearer the end of the descending colon. Hoffmann, Aeby, Luschka, Küttner and Pirogoff all describe this ligament, but most anatomists hold it to be always an artificial production. I found it in the great majority of the bodies of young children, which I examined but only when the mesentery of either the ileum or the pelvic colon was put on the stretch.

In cases of great distension of the intestines it is probable therefore that there is such a fold present and it might then play some part in limiting the/
the upward spread of the peritonitis.

Structure of Pelvic Colon.

The arrangement of muscular coat requires description. The taeniae of the descending colon spread out on the Pelvic colon to form an almost complete longitudinal muscular coat. The postero-external band gradually passes on to the front and unites with the anterior taenia to form a broad band which occupies nearly the whole width of the bowel in its lower portion. The postero internal taenia spreads out in a similar manner on the back; so that in the lower half of the pelvic colon the longitudinal layer of/
11.

of the muscular coat is complete with the exception of a narrow part on each side: here the circular fibres come to the surface and the intestine presents a series of small sacculations. These however disappear and the longitudinal fibres although thicker in front and behind, form a continuous layer all round as the rectum is approached. The vascular and nervous supply of the pelvic colon do not present any feature of special importance.

Position of Pelvic Colon in the Abdomen in the Adult.

The typical position of the pelvic colon in the adult is in the true pelvis as was stated above. According to Jonnesco it is found there in 92% of cases. There is no doubt however that this figure is much too high. Samsom found it in its typical position in only 44% of the bodies he examined. Engel examined 100 cases: he found the pelvic colon in its normal position in 62 of these.

In the other 38 cases the positions was as follows:

Rising upwards into the left hypochondrium. 8%
Rising in centre of the abdomen almost to epigastrium. 6%
Extending to the hepatic flexure of colon. 2%
Lying in front of caecum. 6%
Lying transversely across the lower part of the abdomen just above the symphysis pubis. 16%

\[\frac{38\%}{\text{38\%}}\]
There is therefore practically no part of the abdomen in which one may not find the pelvic colon. According to Engel the reason for this variety of position is as follows.

"The change of position of a hollow abdominal viscus depends on the degree of its distension with gas. The pelvic colon is more liable to gaseous distension than any other part of the intestinal canal." Moreover when the pelvic colon is distended either naturally or artificially with gas, it invariably tends to rise upwards in the abdomen, as an arch. Thus though the true pelvis may be the normal position for the empty pelvic colon, a distended pelvic colon is very seldom found there.

In the child at birth the pelvic colon presents numerous points of difference from that in the adult. It differs from the adult pelvic colon not only in its relative size and length but in its position in the abdomen and in the relative length, in the shape and in the attachments of its mesentery. To properly grasp the nature and the cause of these differences it is essential to understand the development of the pelvic colon and its mesentery. The following description of the development is drawn partly from my/
my own observations and partly from Toldt's work on this subject.

In a six week's old embryo the whole intestinal canal possesses a mesentery. The portion of mesentery belonging to the lowest part of the bowel (the hind gut) has a proportionately long line of attachment in the mid-line of the body.

The mesentery is however short and becomes shorter the farther down one goes till it is completely lost in the pelvis. At the beginning of the third month (traced from below upwards) the bowel rises upwards in the mid-line without any S shaped bend; in the lumbar region it inclines slightly to the left so as to lie on the mesial border of the left kidney. The mesentery is still attached in its whole extent to the mid-line.

In the second half of the third month the first appearance of a sigmoid flexure is seen in the form of a loop of bowel which winds outwards in front of the genital gland into the left iliac fossa and then runs upwards and inwards (lying on the posterior abdominal wall) to the lower pole of the left kidney. The mesentery is still attached to the midline right up to the pancreas and lies free on the posterior abdominal/
abdominal wall.

In the first half of the fourth month the rectum lies in the mid-line of the pelvis, from there the pelvic colon rises upwards directly in the mid-line to the level of the third lumbar vertebra, here it bends outwards to the left under the lower pole of the left kidney where it makes an abrupt loop downwards and then ascends into the descending colon at the outer border of the left kidney. The mesentery is still attached to the mid-line but the part of it belonging to the pelvic colon is raised in a tense suspending fold which is continued into the descending meso-colon. It is this fold which determines the ascending position of the lower part of the pelvic colon which now rises upwards instead of lying lax in the iliac fossa as it did earlier in development. The production of the fold is dependent on the stretching of the general mesentery of the large intestine due to the descent of the caecum downwards and to the right. The free descending meso-colon becomes stretched over the duodenal jejunum flexure and at this point is raised in a fold. The fact that the lower part of the pelvic colon becomes pulled up at this time, shows that such/
At the beginning of the fifth month (sometimes at the end of the fourth) the descending meso-colon is found no longer free and capable of being lifted up as far as the mid-line but is adherent above as far out as the pole of the left kidney. The process of adhesion gradually extends over the front of the left kidney and then outwards directly under the splenic flexure soon to reach the bowel itself. In a downward direction the process of adhesion is not quite regular for a short narrow fold is left on the front of the adherent meso-colon; this is a remnant of/
At the sixth month the pelvic colon has its two typical loops, the lower of which lies in front of the genital gland. The more the pelvic colon is filled with meconium the further it stretches out either to the right over the mid-line or upwards to the hilus of the left kidney.
At the end of foetal life the descending colon is fixed to just below the iliac crest. During the early years of life the process of adhesion still goes on in a downward direction so that what was pelvic colon at birth must later be reckoned as iliac colon.

ANATOMY OF PELVIC COLON

IN THE NEWLY BORN AND IN THE INFANT.

In the middle of the nineteenth century attention was first drawn to the difference in the proportions and relations in the infantile and the adult pelvic colon./
Jacob showed that the large intestine in children is proportionately long. The ascending and the transverse colon (as he pointed out) are comparatively short, the increase in length being to some extent in the descending colon but more especially in the pelvic colon.

Huguir accounted for this by the fact that the large intestine especially the pelvic colon serves as a receptacle for meconium which dilates and lengthens it.

Leichenstern on the other hand says that in the foetus the large intestine grows faster than does the abdomen, and occupies a relatively large part of the abdominal cavity.

Whatever be the cause the fact that the pelvic colon is relatively long in the infant is beyond dispute.

In the adult.

The average length from caecum to iliac colon = 135 c.m.

" " " of pelvic colon = 40 c.m.

(Cunningham.)

In twenty new born children which I examined the measurements were as follows.

Average length from caecum to iliac colon = 31.4 c.m.

" " of pelvic colon = 20.0 c.m.
In sixteen children below two years of age which I examined; -

Average length from caecum to iliac colon = 39.6 c.m.
" " of pelvic colon. = 21.6 c.m.

Thus whilst in the adult the pelvic colon bears to the rest of the colon, as regards length, the relation of 1 to 3.5, in the newly born child the similar relation is 1 to 1.7 and in the first two years of life the relation averages roughly 1 to 2.

I must acknowledge of course that intestinal measurements are always of a somewhat arbitrary nature, as the condition of the bowel as regards distension etc. is not a constant factor in the different cases.

LIMITS OF PELVIC COLON IN YOUNG INFANTS.

Samsom in twenty-three infants below six months of age found that in twenty-two the pelvic colon commenced at the outer border of the left Psoas and ended in the mid-line at the third sacral vertebra when the bowel was empty, but when distended the lower limit rose, sometimes to the level of the sacral promontory.

My own observations fully confirm this statement.

Form.

All observers comment on the marked twining and winding/
winding form of the pelvic colon in infants, as many as three or four loops being sometimes present, and sharp bends and apparent kinks are frequently noted.

Position in Abdomen.

The question as to what is the usual position occupied by the pelvic colon at birth, is one on which much discussion has taken place, and there is great disparity in the records of different observers. Huguier found it usually in the right iliac fossa.

Giralde/
Giralde on the other hand found it much more frequently on the left side than on the right.
(Twenty-four times on the right side in 114 observations.)

Bourcart in 150 new born infants found it on the right side in only thirty-two.

Ssokolow examined a very large number of cases with the following result.

On the right side in 60%.
" " left " " 25%.

In the remaining cases it was in mid-line or partly right and partly left.

In the twenty new born infants which I myself examined it occupied the right iliac fossa in fourteen cases.

In ten children below one year of age I found it in the right iliac fossa in five.

In nine children above one year and below two and a half years of age, I found it five times in the right iliac fossa.

There can be doubt at any rate that the pelvic colon in infants very frequently extends well over into the right iliac fossa especially when it is distended with meconium or gas, and thus in performing colostomy for atresia of the rectum an incision to the right/
right of the mid-line will give almost certain access to the distended pelvic colon. The relatively small pelvis and the relatively large pelvic colon in the infant account for the position.

Bourcart described three positions in which the infantile pelvic colon may be found.

1. Ascending up towards the spleen.
2. Transverse to the right iliac fossa.
3. Descending into true pelvis.

In 150 cases he found the ascending type in 112, the transverse in thirty-two and the descending in only six.

All statistics agree as to the rarity of finding the typical adult position of the pelvic colon in the infant. (in probably not more than four per cent of cases)
Fig. 9
Large Intestine of
unusual length in a newly born
child. The convoluted pelvic
colon is well seen.

Fig. 10
Pelvic Colon
in a newly born infant.
Convoluted and distended
with meconium.

Fig. 11
Pelvic Colon
in a newly born infant.

Fig. 12
Distended Pelvic Colon and
Transverse colon in a child at 2 years
Shows also delayed descent of the Eecum.
PELVIC MESO-COLON IN THE INFANT.

At birth the pelvic meso-colon differs in several respects from the adult type.

The infantile type is proportionately longer, it is much more elastic and its attachments differ considerably from those found in the adult. In my twenty cases the average length of the pelvic meso-colon was 4.1 c.m., the longest being 7.5 c.m. and the shortest 2.3 c.m..

In the newly born the attachment of the pelvic meso-colon to the posterior abdominal wall extends from the outer border of the left psoas muscle at the level of the sacral promontory upwards and inwards to the front of the third or fourth lumbar vertebra, and then downwards in the mid-line (or slightly to the left or right of it) to the level of the 3rd sacral vertebra.

The parietal attachment of the pelvic meso-colon is thus proportionately long in the infant. This extensive attachment is said to account for the extreme rarity of volvulus in young people. The angle of the mesentery is situated very high up (3rd or 4th L. vert.) in the infant and it is larger and blunter than in the adult. (Neter)\textsuperscript{19}

When the pelvic colon of an infant is lifted upwards the inguino-colic fold of peritoneum can usually/
usually be very distinctly seen. With the successive years of childhood the infantile type of pelvic colon and meso-colon gradually merges into the adult type, but occasionally the infantile type may persist through life. Markel describes four types of pelvic meso-colon.

I. Long, highly situated angle, long mesentery and long pelvic colon.

II. Long, highly situated angle, short mesentery and short pelvic colon.

III. Low lying and short angle, long mesentery and long pelvic colon.

IV. Low lying short angle, short mesentery and short pelvic colon.

No. I. is the "Infantile extreme."

No. IV. is normal in adult life.

Nos. II and III are the intermediate types.
MORBID ANATOMY OF HIRSCHSPRUNG'S DISEASE.

The condition of parts is best studied from below upwards. The anus is normal. Fenwick however reported one case where apparently spasm of the sphincter ani was the cause of the trouble; his case is unique in this respect.

The rectum is as a rule normal. In some cases however it has been found very roomy and dilated. Hawkins believes that the dilated colon always ends by a funnel shaped gradual narrowing and that this funnel shaped part is in many cases the upper half of the rectum. Treves in one case found what appeared to be a congenital narrowing of the rectum in almost its entire length. He expressed the opinion that this state of affairs would probably be found in every case, but as far as I can make out it has not been found either before or since his publication.

Generally the dilatation begins just above the rectum but the lower 2 - 3 inches of the pelvic colon may remain unaffected and in one or two reported cases the pelvic colon has been of normal size and the dilatation has been higher up in the colon. As a rule the pelvic colon is found enormously enlarged/
enlarged having the form of a single loop extending upwards in the abdomen and also slightly to the right; in many cases extending below the right costal margin. The posterior surface of the pelvic meso-colon then comes to look forward and vice versa.

Generally the loop of pelvic colon rises completely out of the pelvis but in some cases the lower part of the loop has been found wedged in the pelvis and apparently exerting a valvular action over the upper end of the rectum. In one or two cases the pelvic colon has not extended so far upwards in the abdomen as its size would have led one to expect, but has filled up the hypogastric and right iliac regions. Shortness of the pelvic meso-colon probably accounts for the position of the bowel in such cases.

In the fully developed condition the dilated and hypertrophied bowel reaches enormous proportions so that on freely opening the abdomen it conceals all the viscera (with the exception of the liver) from view. It may have a diameter of from six to eight inches and in many of the reported cases it is described as having the proportions of a man's thigh.

In one or two cases reported as this condition there has been found a localised sac-like dilatation of/
Fig. 13.
Enormous dilatation
and hypertrophy of
the colon in a boy
age 14½ years.
(after Gec.)

Fig. 14.
Enormous dilatation
and hypertrophy of the
pelvic colon in a boy
age 15 years.
(after Morehead)
of the pelvic colon. Probably it were more correct to classify such cases along with intestinal diverticula than as Hirschsprung's disease.

In most cases the dilatation has gradually ceased as the iliac and descending colons were reached but in quite a considerable number the descending colon has been to some extent dilated.

Less frequently has the transverse colon been found enlarged and still less frequently the ascending colon and caecum. At least two cases have been reported however where the maximum of dilatation and hypertrophy was found in the transverse colon and where the pelvic colon was not appreciably affected.

The small intestine invariably appears quite normal in size and structure.

Apart from its great size the most striking feature of the affected portion of bowel is the great thickness and hypertrophy of its wall, the bowel usually having the appearance of a dilated and abnormally thick walled stomach. This increased thickness of the bowel wall is chiefly due to an enormous hypertrophy of the circular muscle coat (see later).

In longstanding cases the bowel wall has a tough leathery and more rigid consistency and in these cases there is found considerable fibrous hyperplasia in the bowel/
bowel wall and evidence of "chronic interstitial enteritis." 

Ulceration of the mucosa has been noted in not a few cases, (at least one case has been reported where death was due to the perforation of such an ulcer), but it is not the rule.

When present the ulcers are generally numerous, irregular in shape and extending down to the circular muscular coat. They are usually found at the most distended portion and have thus been given the name "Dehnungs-Geschwure" (Kredel) Ulceration has been more frequently found in cases where the dilated colon contained hard scybalous masses than in those where the content was almost exclusively gaseous.

The content of the dilated colon varies considerably in different cases.

In the majority of cases it is gaseous with scanty semi-solid faeces. In a fair proportion of cases however the colon has been found filled with a solid mass of hardened faeces weighing many pounds and from one or two cases smaller very hard masses almost meriting the name of enteroliths have been removed.

Microscopic Appearances.

Mucous Membrane. Where no ulceration is present the/
the mucous membrane except that it shows a complete absence of folds may not appear to be profoundly altered. It may have a more or less complete epithelial covering and its normal mucous glands. Almost invariably it shows some degree of leucocyte infiltration, indicating a state of chronic irritation. In many cases however much more marked changes have been found. In some the surface epithelium had entirely disappeared over large areas and only traces of the glandular structures remained.

At the site of an ulcer the mucous coat is entirely wanting, and round the ragged margin of the ulcer great leucocyte infiltration is seen.

Muscularis Mucosae.

In the majority of cases this has been found slightly hypertrophied, in one or two cases greatly hypertrophied and thickened, and in several other cases interstitial fibrous changes are described. In this layer also leucocyte infiltration especially round the blood vessels is frequently seen.

Submucous Coat.

This layer always shows a much denser and more fibrous structure than normal. The connective tissue fibres are thickened, the bloodvessels are more numerous than normal and have thickened walls and round/
round them lymphocytes are collected in great numbers.

Circular Muscular Coat.

This is the coat which almost always is chiefly responsible for the thickening of the bowel wall. It shows an extreme degree of hypertrophy being frequently four to five times its normal thickness. This increase in thickness is found to be due usually to a true muscular hypertrophy the muscle fibres being larger and more numerous than normal.

There is also always some degree of leucocyte infiltration between the muscle fibres and round the bloodvessels. The latter appear to be larger and more numerous than normal.

In old-standing cases thick connective tissue fibres are seen either between or replacing the muscular fibres.

At the site of an ulcer one may see sometimes definite necrosis of this layer.

Longitudinal Muscular Coat.

This shows generally a slight degree of hypertrophy but is rarely more than twice its normal thickness. In one case (Bing) there was very pronounced fibrous degeneration of this coat.

Serous Coat
Serous Coat.

This generally shows a slight degree of fibrous thickening. In Mya's case however the serous coat was enormously thickened being twice as thick as any of the other layers.

Fig. 18. — Coupe du colon valvulae (apres Conroy).
ETIOLOGY.

Probably in the case of no other disease does such a diversity of opinion exist in respect to its etiology as is found in the case of the malady under discussion.

This diversity of opinion among the most competent observers is so great that one is almost driven to the conclusion that the dilatation and hypertrophy of the colon which we know as Hirschsprung's disease may be produced by any one of several factors and that no one cause will ever be found which will account for all the cases which have been reported as examples of this disease. If we adopt the view taken by some writers (e.g. Bing) that, if in a given case any definite obstruction to the onward passage of bowel contents be proved, the case should not be reckoned as Hirschsprung's disease, then we must exclude almost one half of the reported cases.

The most generally accepted view is that if in a case of dilated and hypertrophied colon no distal organic obstruction, such as stricture, growth, or compressing band, be found the case may be reckoned as one of Hirschsprung's disease.
I cannot do better than first give briefly a synopsis of the different views which are held in regard to the etiology of the affection with the names of the supporters of each view, as far as I have been able to collect them.

Theory (1).

"The dilatation and hypertrophy are truly congenital." This view is strongly held by Hirschsprung himself. Mya called the disease "Megacolon Congenito." Concetti calls it "Megalocolie". Kredel describes it as "Eine Art Riesenwuchs des Colon". Generisch, Griffith, Sternimann, and Neugehauer all support this view.

Theory (2).

"That abnormal length and sling formation of the pelvic colon, with or without secondary kinking, volvulus or valve formation, is responsible for the condition."

This view has very numerous supporters among whom may be mentioned: - Jacobi, Broadbent, Morgan, Duhamel, Johanessen, Neter, Frommer, W. Bergmann, Baginski, Mulberger, Bittorf, Delkerkamp.

This may be called the "Anatomical Theory."

Theory (3).

That an abnormally long pelvic meso-colon with secondary/
secondary twisting of the pelvic colon is the cause. This view has not obtained much support but is brought forward by: Borth, Gourévitch, and Schuckmann.

Theory (4).

That there is congenital aplasia of the muscular coat of the intestine with secondary distension. This is the "Ektacolie" of Concetti, Berghniz supports this view.

Theory (5).

That there is a weak tonus of the muscular coat of the colon the result of defective innervation. "Neuropathic dilatation and hypertrophy" Hawkins calls it. Bing is a strong supporter of this view. Lunn, Fitz, and Lennander favour this view.

Theory (6).

That spastic contraction of the lower part of the colon or of the sphincter ani cause dilatation and hypertrophy above. Fenwick, See, Hitchin, Drew and Schreiber bring forward evidence to support this view. Wilms believes "functional spasm of sphincter ani" to be the cause.

Theory (7).

That it may be caused by a "primary inflammatory alteration/
alteration in the wall of the colon." Griffiths is the only advocate of this view.

Theory (8).

That there is congenital narrowing of the rectum or the lower part of the pelvic colon. Treves first advanced this view. Hobbs and de Richmond, and Gruneberg support it in a slightly modified fashion. If this were actually the case the prefix Idiopathic would be absurd.

Theory (9).

That in some cases inco-ordinated muscular action of the colon, similar to that present in congenital pyloric stenosis may be the cause.

(J. Thomson)

I will not attempt here to weigh all the evidence for and against these various theories but will confine myself to the following questions.

I. Is the condition truly congenital?

II. Can it be explained on anatomical grounds?

III. May not another factor which previously has received but scant notice be potent in the production of this morbid condition.

I. Everyone is agreed that the cause of the condition must be a congenital one, but no definite evidence has been brought forward to show that there is an abnormally/
abnormally large colon with thick hypertrophied walls in any of these cases at birth, as Hirschsprung and Kredel believe.

The only recorded case where the signs and symptoms of this disease were present from birth is that of Escherich. The case was that of a child in whom at birth the abdomen was noticed to be distended and where vomiting of meconium shortly followed. The child lived to the age of $3\frac{1}{2}$ months and during this time had a swollen abdomen pronounced constipation and vomiting.

At the post mortem the pelvic colon was very large, long and distended, the transverse colon was distended. Microscopic examination revealed no trace of hypertrophy in the bowel wall. Eschrich was of the opinion that the cause of the trouble here was an abnormally long and large great gut, and that if the case had lived it would have developed into a typical case of Hirschsprung's disease with "Arbeitshypertrophie" etc. In no case where a child has died during the first few days of life has the typical hypertrophy of the bowel wall been demonstrated.

II. THE ANATOMICAL THEORY.

Briefly this is "that an exaggeration of the long/
"long sling like pelvic colon found normally in the infant is the cause of this disease."

Some hold that simply the increased difficulty in the onward passage of the bowel contents caused by the abnormal length and numerous bends in the pelvic colon is sufficient to cause the condition; others think that the abnormal length and multiple bends predispose to kinking and valve like action and dilatation and hypertrophy occur above such kinks. There can be no doubt that in the very long and much convoluted pelvic colon found in some infants any degree of distension would be very liable to cause a certain degree of kinking; should this once happen one can readily understand what the sequence of events would be. The stagnation and gas formation above the kink would only tend to accentuate the latter, and even should the kink for the time-being be overcome, it would just as readily again occur, and the recurrence of attacks of obstruction would gradually lead to dilatation and hypertrophy of the bowel above. The attacks of complete obstruction from which many cases of this disease periodically suffer lends support to the belief that kinking plays a considerable part in their etiology.

It/
It has frequently been urged against this theory that were obstruction from kinking the cause, not only would the pelvic colon be dilated, as it so frequently is in this disease, but the rest of the large intestine would be always found dilated as well. This argument however is not based on fact, as it has now been repeatedly proved that the pelvic colon is the most readily dilatable part of the large intestine. This can be demonstrated experimentally by cutting across the ileum and injecting air or fluid into the lower cut end. It is found that the pelvic colon dilates to a much greater extent than any other part of the large intestine. (See photo)

Moreover apart from cases of this disease altogether, one occasionally finds (in children at least)
least) the pelvic colon greatly distended with gas while the rest of the colon shows little or no dilatation. (see Fig. 17)

Furthermore, Zillner has drawn attention to the fact that the pelvic colon is the seat of election for rupture of the large intestine in new born children. Lastly Sudsuki investigating the subject of intestinal diverticula, found that true diverticula occur more frequently in the pelvic colon than in any other part of the intestinal canal.

That/
That actual kinking does sometimes occur when the disease is established has now been definitely proved on several occasions, more especially by Ibrahim and by Neter.

The situation of the kink or valve may differ in different cases but many believe that it is the pelvic colon which kinks over the upper end of the rectum, valving the latter. In such a case the distension of the pelvic colon above the valve simply compresses the rectum more firmly and aggravates the obstruction, but frequently as the distended pelvic colon rises in the abdomen (which it always does when greatly distended), its lower end is drawn up out of the true pelvis and the valve is opened with at least temporary relief.

Further, in support of this theory is the fact that experience has shown that in some cases of this disease, if a small colostomy opening be made in the dilated bowel, which simply allows gas to escape, not only will the troublesome obstructive symptoms disappear but the bowels will act regularly.

Perthes demonstrated, in a case in which he had performed colostomy for this condition, that though rectal injections readily escaped from the preternatural anus/
amus, injections through the fistula did not come away per anum, showing the present of valvular obstruction.

The mere fact that long sling like pelvic colons are frequently seen in the post mortem room in cases which have had no symptoms of this disease, is in my opinion no evidence against this theory, as the second factor of kinking is always necessary in this class of case.

On the other hand, although we allow that the anatomical structure of the pelvic colon in newborn infants and young children does predispose it to kinking with obstruction, and though we may grant that this anatomical factor does play a considerable part in the causation of many cases of this disease, we/
we must at once recognize that in a considerable proportion of cases this "kinking theory" of causation is definitely excluded. I refer particularly to those cases where a dilated and hypertrophied pelvic colon gradually narrows in funnel shaped fashion into a more or less normal rectum and where a kink or valve was obviously not the causal factor. (Hawkins lays particular stress on the frequency of this class of case) I believe that a physiological factor carried to excess plays a large part in the etiology of such cases if not of all cases.

III. This factor I believe to be primary over-distension of the pelvic colon with meconium.

My grounds for this belief are as follows:--
In examining the abdomensof newly born infants I was struck by the fact that while the small intestine was usually more or less collapsed and empty, the large intestine but more especially the pelvic colon was almost invariably full of and was frequently distended with meconium. In some cases this was so pronounced that the pelvic colon resembled very much in its proportions that seen in Hirschsprung's disease and suggested some distal obstruction, but no/
no kinking or rectal atresia was found in any case.

Evidently at or just prior to birth there is a physiological propulsion of the meconium to the lower part of the large bowel, distending it. The degree of this distension varied greatly in different cases and one could readily imagine that if this physiological distension were carried to excess, the ability of the bowel wall to contract after birth and expel/
Fig. 22
Pelvic Colon in a newborn infant
Showing great distension with necrosis and in appearance suggesting a case of Hirschsprung's disease.

Fig. 23
Section of wall of Pelvic colon figured above. Shows no hypertrophy of the muscular coats.
expel the meconium would be seriously impaired.

Moreover should the tonus of the infant's bowel be below normal, the pelvic colon would be the part where it would most probably dilate as this part is normally distended before or at birth. Abnormally weak tonus is not in my opinion an essential factor however.

Granted that overdistension has taken place at or before birth, one can readily imagine that this overdistended pelvic colon will not empty itself after birth so soon or so readily as normal, just as we find defective action in an overdistended bladder. Now we know that though meconium is sterile at birth, within twelve to twenty-four hours after birth it contains bacteria which cause fermentation and gas formation. This excessively distended pelvic colon will be unusually susceptible to fermentation, and the resultant gas formation will further distend it.

Fig 28
Section of wall of pelvic colon figured on the next page.
There is no appreciable hypertrophy of the muscular coats.
If we remember that we are dealing with an organ in a state of active development when its final form, size and position have not as yet been attained, and when it is peculiarly susceptible to forces such as pressure and distension, I think we will acknowledge this explanation to be a reasonable one.

I consider therefore that we have to deal with a pelvic colon that from the very outset of extra uterine, life must act at a disadvantage, and which/
which never contracts to its normal calibre. This primary over distension has two consequences which are strictly related to and dependent on each other. The one is hypertrophy of the bowel wall, the other is further dilatation.

The hypertrophy of the bowel wall is easily explained. The dilated bowel cannot grip its content and force it on as a normal piece of bowel would, it therefore makes constant and only partially successful efforts to do so; this constant action results in hypertrophy of its muscle wall, the "Arbeits Hypertrophie" of German writers. This imperfect propulsion of content, however, leads to a greater or less degree of permanent stagnation in the colon, with gas formation and further distension, so that a vicious circle is kept up and leads to the enormous hypertrophied colons so characteristic of an advanced stage of this disease.

The relation which the hypertrophy bears to the dilatation determines the fate of the case. Where the hypertrophy fails to keep pace with the dilatation, we get early obstructive symptoms with distension and frequently death, in infancy or early childhood, from toxaemia.

Where the hypertrophy is sufficient to compensate for/
for the dilatation, the child may reach adult life, suffering only from a slightly swollen abdomen and a certain degree of constipation. Adult life being reached compensation does not usually fail till the degenerative changes of old age begin to set in; then, from fibrous changes occurring in the hypertrophied wall, compensation fails, the bowel dilates further, and leads to the well known symptoms of this disease.

In support of this, it is worthy of note that it is extremely rare to meet with a case of this disease appearing for treatment between the ages of twenty-two and forty, although numerous cases have been reported where the patient was over fifty years of age.

One might almost compare the condition to that of a heart which in early life had been so acutely dilated that complete restoration to its normal size was impossible, and in which a regurgitant valvular lesion was also left. (The regurgitation is comparable to the gas formation in colon) By hypertrophy of its wall the dilatation is compensated. If the hypertrophy be not in proportion to the dilatation, troublesome symptoms or even early death will ensue. Should the hypertrophy however exactly/
exactly compensate for the dilatation, the patient may live through many years without any cardiac symptoms. As old age comes on however and fibrous changes occur in the myocardium, compensation may give way and the disease again asserts its presence.

CLINICAL.

For permission to use the following five cases I am indebted to the kindness of Mr. Stiles and Dr. John Thomson.

Case I. was under the care of Dr. Thomson at the Royal Hospital for Sick Children, Edinburgh.

Case IV. was also treated by him in the first place.

Cases II, III and IV were under the care of Mr. Stiles in the same Hospital.

Case V. was treated by Mr. Stiles in Chalmers Hospital.

While House Surgeon in Chalmers Hospital and in the Children's Hospital I was fortunate in having the opportunity to study three of these cases. (Cases III, IV and V.)

The order the cases are given in, is based on the method of treatment each received, this being in successive cases progressively more radical.
Case I.  John M.  Aet. five weeks.

Admitted July 7th 1906.

Complaint.  Constipation and swelling of abdomen since birth. Vomiting for two days.

History.

Child appeared healthy at birth. Bowels did not act during the first three days of life in spite of castor oil. On third day a motion after an injection. During the first three weeks bowels never moved naturally and injections had to be given regularly. For the two weeks before admission the bowels have acted naturally. Motions pale. Has had occasional vomiting ever since birth, but during the past two days it has been persistent, and vomit has been green in colour. The swelling of the abdomen was noticed shortly after birth, and this has persisted and for the past two days has been much more marked than formerly.

Takes nourishment well but is getting very much thinner.

State on Admission. Weight 7lbs. 10 oz.

Small pinched face. Nutrition poor; Abdomen is much distended, the skin tense and glazed and superficial/
superficial veins unduly prominent.

Circumference at infracostal plane 16 inches.

" " umbilicus 15 "

At intervals, intestinal patterns can be very distinctly seen. In the epigastric and hypochondriac regions a large piece of gut stands out prominently in rigid spasm and can be easily grasped in the fingers. Another large and prominent piece of gut can be seen and felt passing from the left side of the umbilical region downwards to the left Iliac region. Other smaller patterns, frequently standing out, can be both seen and felt. In the right Iliac region very marked splashing can be elicited by palpation.

Borborygmi are occasionally heard. Palpation of abdomen between the intestinal spasms reveals nothing.

Nothing abnormal in heart and lungs. Examination under chloroform revealed a somewhat narrow rectum but no definite stricture.

Treatment. Daily lavage of bowel was followed by great improvement. Ten days after admission child became suddenly collapsed and in spite of all treatment died the following afternoon.

Sectio. The whole of the large intestine was dilated/
dilated and hypertrophied, the hypertrophy of the muscular coat being the most striking feature.

The condition was not specially localised to any one portion but affected the whole of the great gut.

No stricture of rectum.

The small intestine appeared if anything slightly thickened.

Pylorus and bladder showed nothing abnormal.

Liver showed cloudy and fatty changes.

Lungs showed some acute Bronchopneumonia.

Fig. 26

Section of wall of colon from Case I

Shows the partial destruction of the mucous coat and its hypertrophy of the circular muscular coat.

(low power view)

Case II.  Lizzie N. Aet. 2 years and 4 months.

Admitted Nov. 3rd 1900.

Complaint/
Complaint. Chronic constipation and swelling of the abdomen.

History.

Ever since birth child has had a swollen abdomen and has been very constipated, the bowels acting every four or five days.

At the age of eleven weeks suffered from an attack of complete obstruction of the bowels, which was diagnosed as intussusception, but no blood was passed per rectum and the condition was relieved by enemata.

During the first year and a half of life enemata were given regularly to secure an action of the bowels. At the end of the period child had a very severe attack of diarrhoea lasting for one week. The motions were very frequent and on several occasions contained blood.

Motions were at all times very offensive and the child daily passed much flatus of very offensive odour.

Fourteen days before admission after a few days of very troublesome flatulent distension child had another attack of diarrhoea. There was again blood in/
in the stools. Child has always had a very large appetite and has otherwise been healthy.

State on Admission.

The abdomen is enormously distended and the superficial abdominal veins are dilated and prominent.

No patterning can be made out. Tympanitic note on percussion all over. No resistance or mass palpable.

Rectal examination entirely negative. Abdomen was examined under chloroform anaesthesia but with negative result.

A long rubber tube was passed up the bowels for twelve inches and the colon irrigated. Two copious evacuations resulted.

On November 6th the abdomen was explored through a median incision below the umbilicus. The entire length of the intestine was examined. The small intestine was normal. The large intestine was dilated throughout its whole extent but especially the pelvic colon and upper part of rectum where there was also some hypertrophy of the bowel wall. No obstruction was found. Abdomen closed. On the night after the operation child commenced to vomit. This continued throughout the following days and she died.
died two days after the operation.

Sectio. No peritonitis.

The colon was found dilated throughout its whole extent, but especially in the sigmoid flexure, and full of faeces. Very little hypertrophy of its wall. No definite organic obstruction anywhere although the rectum in its lower part appeared relatively narrow.

Case III. Herbert Brown - 5½ years.

Admitted to hospital on June 23rd 1902.

Complaint. Constipation since birth.

Swelling of abdomen since a few weeks after birth. Both these conditions have become more pronounced in the past few weeks.

History.

Child was thin and badly nourished at birth. In spite of castor oil and enemata bowels did not act for almost a week after birth. The child was born on a Tuesday and bowels did not move until the following Monday.

During infancy the child constantly required opening medicine and injections especially during teething/
teething. When a few weeks old, mother noticed that abdomen was unduly large and that it became at times very distended.

Diarrhoea on one occasion (when one and a half years old) in summer when it was prevalent in the neighbourhood. From infancy has required enemata at least once a week.

The child has frequently had acute attacks of abdominal distension with constipation, lasting for about a week and coming on and passing off gradually. The doctor who attended the child prescribed castor-oil every morning and in addition the iodide of iron and cod liver oil, and an open air life, as the diagnosis at this time was Tuberculous Peritonitis.

The swelling of the abdomen however remained and except during a few weeks at the end of 1901, the child was as constipated as ever. In March 1902, the child had an unusually severe attack of obstipation and distension, the legs being swollen on this occasion.

This subsided however under treatment with castor oil and enemata.

In the end of April 1902 a similar attack of distension which on this occasion did not yield so/
so completely to treatment. The swelling diminished only slightly under treatment, with enemata and two weeks before admission, after a chill from sitting with wet feet, the distension again became much more marked. In spite of injections the abdomen increased in size every day, the child refused food and lost flesh rapidly.

On the day before admission passed a great quantity of flatus and got much relief but this was only temporary, the distension increasing rapidly.

**State on Admission.**

An anaemic, thin and feeble child. Pulse rapid, small and weak. The abdomen is enormously distended and dilated, superficial veins can be seen coursing over it. The distension appears greatest in the upper part of the abdomen and the lower part of the thorax is broadened out.

No peristaltic waves can be seen. The skin of the abdominal wall is tightly stretched and shiny. On palpation no special resistance can be felt. On percussion general tympanitic note.

On June 24th, 1902 an exploratory laparotomy was performed. On opening the abdomen what looked like an enormous cyst presented. This was found to be a greatly distended pelvic colon with markedly thickened walls.

The descending colon and to a less extent the transverse colon were also distended.

The pelvic colon was brought into the abdominal wound, with five loop sutures of silk, the wall of the bowel was sutured to the muscles of the abdominal wall. A continuous silk suture was now put in uniting the bowel wall to the skin.

![Image]

**Fig. 27**
Photo taken before the bowel was opened.
Shows the great abdominal enlargement, and emaciated condition of the child.

Four days later the bowel was opened and the mucous/
mucous edges stitched to the skin. On opening the
gut an enormous quantity of flatus escaped followed
by a considerable quantity of fluid faeces, the
abdomen flattening out considerably. For some days
after this the child was very weak and lay in a
curious listless and apparently toxaemic condition.
After this however he got rapidly stronger and went
home thirteen days after the operation taking full
diet.

Child remained in good health, bowels acting
regularly through the colostomy opening. In February
1906 child re-admitted as parents very anxious that
the colostomy opening should if possible be closed.

Exploratory incision made in mid-line. The
parts were fully examined. No stricture or organic
obstruction of any sort to be made out but only an
evermously distended and hypertrophied colon.
Abdomen closed without anything further being done,
as it was thought wiser to leave the colostomy
opening.

Seen in June 1906 and again in July 1907 patient
was in the best of health. One motion daily after
breakfast through colostomy opening.

Attending school. Always a good appetite
for/
for food. Abdomen still abnormally large. (See photo)

![Photo of a person with a large abdomen.](Fig. 28)  

*Photo taken 5 years after operation.*  
Abdomen still somewhat enlarged, but general condition excellent.

**Case IV. William M. Aet. Six years.**

Admitted to hospital on March 24th, 1906.

**Complaint.** Constipation from birth. Swelling of the abdomen for the past twelve months.

**History.**

Ever/
Ever since birth child has suffered from most obstinate constipation. No special prominence of the abdomen was noticed till one year before admission. At this time the abdomen began to gradually enlarge and the constipation became more pronounced than ever.

Motions were small, hard, and passed with a good deal of straining.

During the year before admission the abdomen has steadily increased in size and the child has lost flesh and become dull and apathetic. There has been no pain in abdomen at any time.

State on Admission.

Patient is a small, pale and delicate looking child. Nutrition very poor. Tongue dry and furred. Thorax is narrow above but greatly expanded in its lower part.

There is great enlargement of the abdomen especially in its upper part.

Circumference at infra-costal plane 23 inches.

" " umbilicus 22 "

Every now and then in either flank and across the upper part of the abdomen a swelling 2 - 3 inches broad stands out for a few moments and then fades away/
away. This swelling when present feels hard otherwise there is no resistance or mass palpable in abdomen.

Percussion reveals an enormously distended colon filling the abdomen except for a small area round the umbilicus.

The liver is pushed upwards well under the costal margin.

For six weeks the child had treatment with frequently repeated enemata and doses of purgative medicine with no improvement as regards the abdominal distension.

On April 24th 1906,

Circumference at infracostal plane. $24\frac{3}{4}$ inches

" " umbilicus $23\frac{1}{2}$ "

Surgical interference was decided on and on May 10th 1906 the abdomen was opened in the mid-line below the umbilicus. A greatly distended pelvic colon very thick walled and fully 5" in diameter appeared and was with difficulty delivered through the wound.

The large intestine was examined throughout its whole extent. The caecum was of practically normal calibre/
calibre and its wall was not thickened. Ascending and transverse colons normal but descending colon greatly distended and pelvic colon more so, especially in its lower part. A lateral anastomosis was now performed between the ileum (8 - 10 inches above Ileo-caecal valve) and the dilated pelvic colon.

The pelvic colon was first punctured and a great quantity of gas allowed to escape. The site of puncture was made the centre of the anastomosis.

An iso-peristaltic anastomosis 2½ inches long was made.

It was definitely ascertained that there was no organic obstruction below the dilated colon.

For the first week after the operation there was troublesome vomiting; after that convalescence was uninterrupted and child was discharged six weeks after the operation. Bowels moved naturally once in four to five days the motions being pale and semisolid.

An occasional enema still required.
Remained in good health till Aug. 2nd 1906 when he was re-admitted with symptoms of obstruction, vomiting and distended abdomen.

On admission/
On admission:-

Circumference at infracostal plane 27 inches.
" " umbilicus 26\(\frac{3}{4}\) "

Numerous large enemas were given and the symptoms subsided and child was discharged in four weeks.

Seen again in September 1907. For the past year has been in good health. Bowels have acted regularly and there has been no tending to obstructive attacks or distension.
Case V. James D. Aet 60 years.

Admitted to hospital on May 2nd 1906.

Complaint. Great pain, distension of abdomen and constipation.

History.

Except for occasional constipation patient was fairly healthy up till the age of 44. Then he began to suffer from pronounced constipation which has steadily got worse during successive years. Along with the constipation came also distension of the abdomen, which has varied much in degree from time to time, but has always shown a tendency to become more and more marked. At first ordinary aperient medicines gave relief, but latterly he has frequently required enemas to produce a motion.

Ten years ago he had an attack of acute abdominal distension with severe colicky pains and obstinate constipation. This attack lasted for three days but was ultimately relieved by repeated enemata.

During the past four years the distension and constipation have been much more pronounced and he has frequently noticed the bowel standing out in spasm and has been much troubled with rumbling noises in the abdomen. Sometimes the sudden passage of a very large/
large quantity of flatus relieved the abdominal distension.

One month ago he had another attack of complete stoppage of the bowels with great abdominal distension. He was admitted to Chalmers Hospital and treated with enemata. At first he was given an enema of over six pints of soap and water. This was retained for several hours, and only after giving another enema of several pints was the fluid returned with a great quantity of flatus and his symptoms relieved. He was discharged in a week "relieved". For the past week there has been no proper action of the bowels but he has passed quantities of flatus and mucus occasionally. The abdomen has become greatly distended and now interferes with breathing. He has also had frequently recurring spasms of abdominal pain during which the bowel stands out prominently.

State on Admission.

Patient is a somewhat thin but fairly healthy looking man.

Every few minutes he gets an acute spasm of abdominal pain which lasts for about two minutes at a time.

The abdomen is very markedly distended resembling a full time pregnancy, the skin is tense and shiny and/
and the superficial veins are prominent.

Tympanitic note all over abdomen except in hypogastrium (distended bladder).

Rectal examination reveals nothing abnormal except a somewhat enlarged prostate. Bladder emptied by catheter. Enema given, returned without flatus.

Operation May 2nd. Mr. Stiles.

An incision 3½ inches long made ½ inch outside the outer border of right rectus muscle.

When the peritoneum was opened very much distended large intestine presented. With a continuous silk suture a portion of this distended bowel was sutured into the wound, the stitches taking up the peritoneum and the internal oblique and transversalis muscles. A trochar and canula was now plunged into the exposed portion of bowel and a great quantity of gas and fluid faecal matter allowed to escape. With a knife the opening made by canula was now enlarged and a rubber tube was inserted and stitched to the bowel by two silk worm gut sutures. Skin incision now closed with silk worm gut, one stitch fixing the rubber tube.

After the operation great quantities of flatus and fluid faeces passed through the tube into a bucket by/
by the side of the bed, and the abdomen became much reduced in size.

He continued to improve for two weeks when it was thought advisable to explore in the mid-line to ascertain the cause of the obstruction.  

2nd Operation May 15th 1906.  

Incision eight inches long in mid-line below the umbilicus. On opening the peritoneal cavity a very much dilated and hypertrophied pelvic colon was found and it was seen that it was this portion of bowel which had been opened into at the first operation.

The bowel was very thick walled resembling a great hypertrophied stomach, it showed a slight degree of volvulus. This was untwisted.

No organic obstruction of any kind could be found to account for the obstruction symptoms. It was thought wiser to leave the colotomy opening, so the abdomen was closed.

During the next three weeks the patient's general condition improved considerably. The lower bowel was thoroughly cleaned out by enemata and on June 15th pat. was discharged the bowels acting partly per rectum but chiefly through the colotomy opening in the right iliac region. After returning home the/
the colotomy opening gradually closed and by the end of July 1906 was completely healed over.

For the next five months patient remained in tolerably good health, except that occasionally he was troubled by flatulence and rumbling noises in the abdomen. The bowels acted regularly as a rule but he had several slight attacks of diarrhoea.

On Jan. 3rd 1907 he again noticed that his abdomen was becoming distended and obstinate constipation set in. On Jan. 4th he again began to suffer from cramping pains in the abdomen, he passed no flatus and became more distended. On Jan. 5th he was given a large enema but it was returned without flatus. The spasmodic pains now became much more frequent and more intense and the distension was so great as to interfere with his breathing. Repeated enemata were given but without result, and on Jan. 6th patient was again admitted to Chalmers Hospital in a condition of great distress.

State on Admission.

Patient has good colour, lies flat in bed, breathing somewhat rapidly.

Abdomen. There is great abdominal distension, the percussion/
percussion note is tympanitic all over.

Every few minutes pat. has a spasm of pain, when patterning of the abdomen becomes very well-marked. A large coil about 4 - 5 inches, in breadth rising in the left Iliac fossa crosses obliquely upwards and to the right just below the umbilicus into the right hypochondriac region, it then turns across the epigastric region into the left hypochondrium, then down into the left lumbar region where it became lost.

These spasms last from 20 - 30 seconds and then
the abdomen becomes even and globular again. The liver is considerably displaced upwards. The heart dulness is also abnormally high. Immediate operation was decided on.

Operation. 6th Jan. 1907. Mr. Stiles.

An incision five inches long was made through the left rectus muscle below the level of umbilicus. When the peritoneal cavity was opened some clear brownish fluid escaped, and distended colon bulged into the wound. The proximal (smaller) part of the loop of pelvic colon was the first to come out through the wound. It was about three inches in diameter. The wound was now slightly enlarged and a hand introduced into the abdomen. The larger or distal end of the loop was found to be adherent at one point to the old colotomy cicatrix. By putting slight traction on the loop just above this point the whole of the distended coil proximal to this point was delivered through the wound. The distal end of the loop was enormously hypertrophied and distended, resembling in size a man's thigh and being quite 5 - 6 inches in diameter.

The abdominal cavity having been shut off with sterile swabs, the bowel was punctured with a trochar and/
and canula. A great quantity of flatus and some liquid faecal matter escaped and the loop completely collapsed. It was decided to resect this loop of bowel and do an end to end anastomosis. This was accordingly done. The difficulty occasioned by the disparity in size of the two ends of the loop was got over by partially closing the large distal end till it just fitted the small proximal end, just as was formerly done in a Billroth's partial gastrectomy.

The abdomen was closed in layers by three rows of sutures.

The patient stood the operation well his pulse at the conclusion being 74, strong and regular.

For the first two weeks after the operation patient very did well. The stitches were removed on the tenth day/
day and the wound had healed by first intention. On the twelfth day after the operation however he developed cystitis, (he had suffered from this on two previous occasions) and had much straining with micturition. On the fourteenth day after the operation after straining to pass water, the abdominal wound was found to have completely opened up and had to be stitched, under an anaesthetic.

Convalescence was somewhat protracted owing to the cystitis.

Patient also suffered during the first four or five weeks after the operation from spasmodic griping pains in the portion of the large pelvic colon which had been left behind, and during a spasm this could be seen standing out in the hypogastric region.

The frequent passage of a stomach tube per rectum brought away large quantities of flatus and gave most relief to this condition. Numerous drugs were tried for the chronic flatulence and spasmodic pain. Opium and calomel given together and later B naphthol were found to give most relief.

After a somewhat trying and tardy convalescence patient went home ten weeks after the operation.
The bowels were now acting regularly and all obstructive symptoms had disappeared.

Before sending patient home an examination of the lower portion of the colon was made with Mummery's Sigmoidoscope. It was easily introduced 28c.m. It was ascertained that no ulceration was present.

After discharge patient remained well for two weeks the bowels acting regularly. Then constipation again set in, with swelling of the abdomen, and the old griping pains. Repeated enemata were of no avail and on April 3rd 1907 patient was again admitted to Hospital.

State on Admission.

Patient looks ill and skin has a yellowish tinge. There is marked distension of the abdomen in its lower half the dilated colon standing out clearly.

Fig 32
Diagrammatic sketch of abdomen on admission on April 3rd 1907 - abdomen during peristaltic spasm.
Every now and then a spasm comes on and the patterning of the abdomen is then more distinct. Per rectum. Through the anterior wall of the rectum a tense rounded swelling can be felt completely filling up the pelvis.

A long tube was passed per rectum but no flatus came away.

A turpentine enema gave no relief. At 2 p.m. on the 4th April a hydrocele trochar and canula was plunged through the abdominal wall, at the site of the old colotomy wound, into the distended colon. Gas immediately escaped and gave patient relief. The canula was left in situ being fixed with strapping.

On April 5th the spasms had returned and patient showed signs of peritonitis, therefore operation agreed on.

Operation. April 5th. Mr. Stiles.

Incision made through the old colotomy cicatrix in the right iliac region. When the peritoneum was opened some brownish serous fluid escaped. It was found that that old adhesions in this region had been stitched and the colon was not in close apposition with the abdominal cicatrix. The canula had/
had thus slipped out of the colon and caused slight peritonitis. A drainage tube was introduced down into the pelvis and the wound partially closed. A second incision two inches long was now made in the mid-line directly over the colon and through the old median cicatrix.

With a curved needle and strong silk a portion of the distended colon was sutured to the edges of this wound, thus isolating a portion of the bowel for puncture. A trochar and camula was now plunged into the bowel and a large quantity of flatus escaped. Camula now withdrawn and a small rubber tube inserted through the small opening in the bowel and fixed with a stitch.

This gave relief for a few days, then the opening into the bowel was enlarged and the mucous membrane stitched/
stitched to the skin edges of the wound so as to give patient a permanent colostomy opening at this point.

There was great improvement in patient's condition after this and he went home three weeks later in good health, the bowels acting chiefly per rectum but also slightly through the small colostomy opening.

Patient returned in October 1907 to report. "Says he is in better health than he has been in for years; bowels act regularly per anum. No faecal matter comes through the colostomy opening which simply acts as a safety valve, allowing flatus to escape occasionally. The colostomy opening causes him very little inconvenience and he is eminently satisfied with his condition."

Pathological report on resected portion of bowel.

The bowel is very much dilated and shows great hypertrophy of its walls. There is no ulceration. Microscopically. The increase in thickness is found to be due almost entirely to an enormous hypertrophy of the circular muscular coat.

This is about four times its normal thickness and has the appearance of a pure hypertrophy, the muscular fibres being larger and more numerous than normal. There is a slight increase in the intestinal fibrous/
fibrous tissue between the muscular bundles but this is not a striking feature.

The longitudinal muscular coat is slightly thicker than normal.

The mucous and submucous coats show some slight increase in fibrous tissue but are otherwise normal.

The question now at once arises as to whether this was really a case of Hirschsprung's disease or whether the condition, appearing as it did in a man over 40 years/
years of age, was not simply the result of chronic constipation and slight recurring volvulus.

Personally I think it must be regarded as a true case of Hirschsprung's disease which had been completely compensated in early childhood but in which with advancing years compensation gradually became inadequate, and led to distension with secondary kinking and slight volvulus, with the resultant obstructive symptoms. The volvulus, which was slight, was I feel sure, a secondary development dependent on the great distension of the pelvic colon.

In this case also there was a certain amount of kinking of the lower part of the pelvic colon over the upper end of the rectum, as was shown by the return of the obstructive symptoms after the proximal part of the pelvic colon had been resected.

The case is instructive from the point of view of treatment, as showing the value of a small colostomy opening, as a safety valve, in cases where the affected portion of bowel cannot be completely resected.
Signs and Symptoms.

The clinical picture presented by the disease is a very typical one, but it varies according as to whether the compensatory hypertrophy of the bowel is sufficient to overcome the dilatation or not. Thus we find cases which run a more or less acute downward course, compensation being quite inadequate.

Others again are so well compensated that they present few and slight symptoms, whilst in others compensation is not quite sufficient and they run a chronic course with every now and then an attack of more acute symptoms, and in any one of those attacks a fatal issue may ensue.

There are certain symptoms however which are present in greater or less degree in all cases, and which taken collectively in a young person are almost pathognomonic of this disease. I shall consider these first.

Constipation.

This is characteristically present and almost invariably dates from birth. Generally it is of a very pronounced type, and it is not uncommon to get a history that the bowels have not acted for several weeks. In very many recorded cases there was/
was no motion for four to five days after birth in spite of purgative medicine given by the mouth, and an enema was required to procure a motion.

This constipation persists, the child constantly requiring opening medicine and in many cases never having a natural motion but always requiring an injection. When the child is weaned (this has been repeatedly noted) the constipation becomes still more pronounced. A feature of the constipation is that it does not yield as a rule to purgative medicine given by the mouth, but much more readily to a large rectal injection.

In some cases the insertion of a small piece of soap per anum has procured a motion. In spite of a very marked degree of constipation the stools may not be hard and scybaloous but as a rule are small soft and semisolid and frequently very offensive.

In some cases however a great quantity of hard scybaloous faeces may collect in the colon, and small hard masses may be passed from time to time.

Flatulence and Abdominal Distension.

Some degree of this is always present. The distension of the abdomen is generally noted shortly after/
after birth, usually within the first three to six months but it may not be sufficient to attract attention till much later. 

In only one case (Escherich) has a swollen abdomen at the time of birth been noted, by a competent observer, although in three other cases, those of Hobbs and de Richemonde, Mya, and Futterer and Middeldorpf there was a history of abdominal enlargement at birth.

Duval collected 29 cases where the time of appearance of the abdominal distension was noted.

<table>
<thead>
<tr>
<th>Time</th>
<th>Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>In the first few days of life</td>
<td>12</td>
</tr>
<tr>
<td>&quot; &quot; &quot; month</td>
<td>2 &quot;</td>
</tr>
<tr>
<td>&quot; &quot; &quot; six months</td>
<td>6 &quot;</td>
</tr>
<tr>
<td>&quot; &quot; &quot; year</td>
<td>4 &quot;</td>
</tr>
<tr>
<td>&quot; &quot; second year</td>
<td>1 &quot;</td>
</tr>
<tr>
<td>&quot; Infancy.</td>
<td>4/29 &quot;</td>
</tr>
</tbody>
</table>

The degree of the distension may be enormous and may seriously interfere with respiration. In practically every case of any standing it is a very striking feature. Borborygmi are often a troublesome symptom, in one case they were so loud as to be "heard all over the house". Many cases constantly/
constantly pass flatus, in others large quantities of flatus are passed at intervals. The flatus is usually very offensive and may create a positive nuisance. (Kredel) Patterning of the abdomen is another symptom (see later). Nutrition is always impaired, the patients are always spare and frequently emaciated.

Symptoms in a case running an acute course.

Here the picture resembles in many respects that of a fairly acute intestinal obstruction but the patient may survive for many weeks.

Constipation is extremely obstinate, nothing short of enemata procuring an action of the bowels and even injections frequently failing to bring away anything but a little flatus. Such a case differs from one of acute intestinal obstruction in that a little flatus comes away from time to time and this fact is often helpful in the differential diagnosis. Respiration becomes rapid and laboured from the upward pressure on the diaphragm.

Vomiting may be a troublesome feature but may be absent altogether. Seldom if ever does it become faecal in character.

The patient emaciates rapidly has a sallow poisoned/
poisoned look and is often drowsy and listless from toxaemia. In some cases there are spasmodic attacks of cramping abdominal pain, suggesting some obstruction such as a volvulus or kink, and at these times the enlarged colon may stand out in rigid spasm.

In such cases if the condition is not relieved by passage of a long rectal tube or by operation death results from toxaemia, or from the perforation of an ulcer. (Dehnungs Geschwüre.)

In some cases Acute Colitis supervenes with profuse diarrhoea and a rapid fatal issue.

Symptoms in Chronic Type.

In this class of case one finds chronic constipation, abdominal distension, and great flatulence but there is seldom any vomiting and the patient is able to go about. He has usually a sallow appearance and suffers from fits of lassitude, the result of constant absorption of toxins from the colon. Appetite for food generally remains good, sometimes indeed is abnormally large, but nutrition is always poor.

Regular rectal injections may be required to procure a motion. Large quantities of flatus are passed from time to time. There may occasionally be/
be a little blood in the stools indicating that some ulceration is present.

Such a case is very apt at any moment to develop the symptoms of the more acute type, and sooner or later, from degeneration occurring in the hypertrophied muscular coat compensation gives way, obstructive symptoms set in and death results from toxaemia. Such cases seldom reach adult life.

Compensated Cases.

In this class the only symptom is the chronic constipation. The abdomen is slightly enlarged but does not attract particular attention. Such a case may improve during childhood and live a tolerably healthy life till the age of 40 or 50 years. Then compensation beginning to fail, the symptoms of either of the above mentioned types may set in and may carry off the patient.

Physical signs in a well-developed case.

The great abdominal enlargement is the most striking feature. The shape of the abdomen is characteristic. It appears greatly lengthened vertically when contrasted with the shortened thorax, and the swelling appears to be, and as a rule actually is/
is greater in its upper than in its lower half.

The greatest circumference of the abdomen is found at the infracostal plane and the lower ribs are raised and pushed outwards, the costal angle being greatly widened out. The abdominal contour is not uniform and rounded in many cases, but there is special bulging and prominence on the left side below the umbilicus.

Considering the size of the abdomen its anterior wall is often remarkably lax, indicating the chronicity of the enlargement. In cases of the acute type however the skin may be stretched and tense and dilated superficial veins may be seen.

Intestinal patterns are often visible as the dilated colon goes into peristaltic spasm. Usually a great loop can be made out extending from the left iliac fossa upwards and to the right into the right hypochondriac region. In very chronic cases the patterning is often much less definite and takes the form simply of a slowly occurring change in the contour of the abdomen.

The percussion note over the dilated colon is generally tympanitic, as the intestinal content is almost /
almost entirely gaseous, in rarer cases it may be
dull however as in them the bowel is distended with
solid faecal matter.

The liver dulness is often displaced upwards to
a marked degree, likewise the cardiac dulness.
Rectal examination reveals the absence of any organic
obstruction there. Sometimes the rectum itself is
large and roomy, in other cases it may be compressed
by a distended portion of pelvic colon lying in front
of it in the true pelvis.

Spasm of the sphincter ani has been described
but is certainly very exceptional.

Swelling and oedema of the legs has been noted
in one or two cases and even slight general cyanosis
the result of impeded and restricted respiratory
action has been observed.

Albuminuria has been noted in several cases,
and in one case haematuria occurred regularly when
the colon was more than usually distended. In this
last case the emptying of the greatly distended colon
by the passage of a rectal tube was always immediately
followed by the passage of large quantities of urine;
evidently a condition of hydrenephrosis from pressure
on the left ureter was present.
Fig. 36
Hinchey's Disease in a boy aged 3 years showing typical abdominal distension (after Roffth).

Fig. 37
Hinchey's Disease in a boy aged 16 years, showing colon standing out in rigid abdomen (after Welch).

Fig. 38
Hinchey's Disease in a man aged 26 years. He had been constipated from birth. The colon in this case was full of solid faecal matter.
Fig. 39

Hirschsprung's Disease in a child 10 months old. Death from inanition.

(after Baginski)

Fig. 40

Enormous abdominal distension in a case of Hirschsprung's Disease.

(after Wallek & Griffiths)
Diagnosis.

In a fully developed case the diagnosis does not present much difficulty, except in a case where the patient is seen for the first time during an obstructive attack. Then the case may closely resemble one of acute intestinal obstruction from any of the well known causes. A careful enquiry into the previous history of the case and the passage of a long rectal tube will however do much to clear up the diagnosis. The difficult cases to diagnose are the slight, fairly well compensated ones, where there is no marked abdominal enlargement and no intestinal patterns are to be seen. This class of case is seldom correctly diagnosed and this is unfortunate as it is here that much improvement may be expected from medical treatment.

Such cases have been confused with the enlarged abdomen of Rickets, with tuberculous peritonitis, (this happened in one case of my series) and even with dilated stomach, to say nothing of the cases which have been for long periods vainly treated with purgatives as ordinary cases of chronic constipation.

A very important aid to diagnosis in such cases is the injection of fluid through a tube passed high up/
up into the pelvic colon when the abnormally large capacity of the latter is disclosed.

**Prognosis.**

This of course varies very much according to whether the case presents itself for treatment in the mild and moderately compensated stage of the condition or in the advanced stage with an enormous colon.

Taken as a whole the prognosis in this disease is a gloomy one and the death rate is very high. Some writers put it as high as 85 per cent. In any case there can be no doubt that a child suffering from the condition lives as it were on the edge of a precipice, and is liable to be carried off rapidly by one of several conditions, chief among which are: acute obstruction, acute colitis, and pneumonia; and should it escape these it may die from slow toxaemia and wasting.

52 Concetti collected thirty cases with a view to finding out the prognosis in this disease. In nine of these cases the result of treatment was unknown as the cases were altogether lost sight of. Of the remaining twenty-one cases, nineteen died, two lived.
<table>
<thead>
<tr>
<th>Age at Death.</th>
<th>Cause of Death.</th>
</tr>
</thead>
<tbody>
<tr>
<td>8 died in first year of life</td>
<td>Acute colitis 10 times</td>
</tr>
<tr>
<td>3 &quot; between 1 and 2 years</td>
<td>Cathexia &amp; wasting 4 &quot;</td>
</tr>
<tr>
<td>2 &quot; 2 &quot; 4 &quot;</td>
<td>Post operative 2 &quot;</td>
</tr>
<tr>
<td>3 &quot; 10 &quot; 13 &quot;</td>
<td>Pneumonia 1 &quot;</td>
</tr>
<tr>
<td>1 was 23 years old.</td>
<td>Cause unknown 2 &quot;</td>
</tr>
<tr>
<td>2 were over 50 years.</td>
<td></td>
</tr>
</tbody>
</table>

Duval also analysed thirty fatal cases.

<table>
<thead>
<tr>
<th>Age at time of death.</th>
<th>Cause of Death.</th>
</tr>
</thead>
<tbody>
<tr>
<td>In first 2 days of life 2%</td>
<td>3 acute</td>
</tr>
<tr>
<td>&quot; 6 months 9%</td>
<td>Obstruction 11 chronic</td>
</tr>
<tr>
<td>Between 1/2 - 1 year 7%</td>
<td>Acute colitis 10</td>
</tr>
<tr>
<td>&quot; 1 - 5 years 23%</td>
<td>Surg. Interfer. 4</td>
</tr>
<tr>
<td>&quot; 3 -15 &quot; 11.5%</td>
<td>Pneumonia 1</td>
</tr>
<tr>
<td>In later years 16%</td>
<td>Asystole 1</td>
</tr>
<tr>
<td>Cause unknown 2</td>
<td>Cachexia 1</td>
</tr>
</tbody>
</table>

It is thus evident that the time of greatest danger to life is the first few years of life, and that if a case can be safely tided over those early years its chances are much better. The explanation of this fact probably is that the child with sufficient compensatory muscular hypertrophy of the bowel passes safely through those dangerous years, and compensation, once/
once established, remains good.

In addition, however, as Hirschsprung himself pointed out, some cases may improve with the advance of years on account of the normal process of development of the large intestine, particularly the fixation of the caecum and the drawing in of the pelvic colon by the shortening of its mesentery.

There can be no doubt of this that with the greater knowledge we now possess of the pathology and treatment of this disease, a better prognosis can now be given than would have been, say ten years ago.

I have collected thirty-seven cases from the more recent literature on the subject which bring out this point.

Of these 37 cases - 19 died.

16 of them received medical treatment only.

21 " " underwent a surgical operation.

Of the 16 cases medically treated 11 died.

2 were completely relieved of symptoms.

1 was partially " " "

1 remained in statu quo.

1 was lost sight of.

Of the 21 cases treated surgically 8 died.

12 were completely relieved of symptoms.

1 was partially " " "

1
This shows a death rate of just over 51 per cent, a much more hopeful aspect than that presented by the older statistics.

A survey of literature has led me to the following conclusions with regard to prognosis. If the case be a young child, if the abdomen be not much enlarged and if the constipation can be controlled by rectal injection, then under suitable and persevering treatment the prognosis is fairly good. Such cases were called by Marfan "Congenital constipation". Graanboom considers them as a separate disease which he calls "Pseudo-megalocolon congenitum" and says that the prognosis is good in such cases without operation.

If in a young child the condition be well established, especially if it have advanced rapidly, the prognosis is bad, as such children are very apt to die of toxaemia, of a sudden acute attack of colitis with profuse diarrhoea, or of some lung affection.

Most of the pronounced cases in young children die in spite of treatment.

If the child has survived till seven or eight years of age then the prognosis improves considerably, for/
for in such a case the compensatory hypertrophy must be more or less adequate.

In older people the prognosis is also fairly good, provided suitable treatment is carried out.

All cases however are liable at any time to the dangerous complications above mentioned.

**Treatment.**

The treatment of this condition is notoriously difficult as is evidenced by the striking diversity of opinion regarding it expressed by the different writers on this subject. It is generally recognized that to cure the condition is, in the great majority of cases, impossible, and our aim therefore must be to decide which are the most successful methods of palliative treatment, that is to say, which methods are the most efficient in obviating the obstinate constipation with its accompanying fermentation, toxaemia, and occasional obstructive attacks.

Can the condition be cured or palliated by medical means alone or is surgical interference called for in all cases?

What are the best lines of medical treatment?

What are the different methods of surgical treatment available?
available for this condition and which of these is most likely to give lasting benefit?

These questions must now be considered, and I shall take them up in the above order.

Treatment, Medical or Surgical?

The great majority of writers on this subject agree that when this disease has reached the advanced stage that it usually has before a correct diagnosis is made, medical treatment alone is insufficient. We must remember however than in some cases great benefit has resulted from medical measures alone, and also that surgical interference in these cases is by no means free from risks, the death rate after operation still being comparatively high. I think it is therefore only right that in the first instance medical means be given a fair trial and only when these fail to effect improvement should an operation be undertaken.

Moreover as the disease becomes more widely known, it will be much more frequently recognized in its early stage, when treatment of a purely medical nature is of undoubted value.

On the other hand I can find no record of an actual cure by purely medical measures in a case of this/
this disease, though several cases successfully palliated by such measures have been recorded.

The only completely successful results, (and they are yearly increasing) have been obtained by surgical treatment, and this fact must be kept in view when advising what therapeutic measures should be adopted in any given case.

Medical Treatment.

This must always have two main objects in view.

(1) To improve the muscular tone of the colon

(2) To prevent stagnation and fermentation in the colon.

The measures which claim attention under this head are dietary, treatment by drugs, lavage of colon, massage, electricity.

Diet. In the first place should this condition be diagnosed in a young infant fed on the breast, weaning should be delayed as long as possible, as a change to artificial feeding has, in every recorded case at this age, been coincident with an exaggeration of the constipation and abdominal distension.

In older children and in adults care must be taken in their dietary to avoid all articles of food which are known to induce flatulence. Most writers recommend a diet that will leave as little residue as is possible. This latter point is however open.
is open to question as it is not unlikely that the large colon could contract on, grip and propel onwards a bulky residue better than it could a small one.

More experience is required however before we can speak dogmatically on this point.

**Treatment by Drugs.**

The value of giving the ordinary purgative medicines by the mouth is doubtful, and some writers go so far as to say that ordinary purgatives are contra-indicated. Those drugs which act by increasing the muscular tone of the bowel are certainly the ones most likely to be of value, and thus Belladonna and strychnine given singly or together have been found useful.

Cheadle has reported numerous cases of chronic constipation in children (many of them had distended abdomens and were probably mild cases of Hirschsprung's disease) in the treatment of which ordinary purgatives had been found valueless but which were all curable or at least palliable under treatment with tonics increasing intestinal tone.

There is singularly little mention in the literature of the administration of intestinal antiseptics or antifermentative drugs. In case V. of my series B Naphthol, Sodium Sulphocarbamate, and/
and charcoal were tried, in no case however with much benefit.

Lavage of the Colon.

This is unquestionably the medical measure of most value. It should be carried out at least once daily. A long rubber tube should be passed per rectum right up into the dilated colon and the latter irrigated with several pints of water. Fermentation and distension are thus to a great extent obviated with benefit in both the local and general conditions of the patient.

In those cases in which the colon is found full of solid faecal matter, warm olive oil enemata frequently repeated are indicated, until the bowel has been emptied, when the daily lavage may be substituted.

Massage.

Abdominal massage has been found to be beneficial in some cases. It is of value not only because it stimulates muscular contraction of the colon, but also because it enhances the strength and tone of the abdominal muscles which are over stretched and weak in these cases. Escherich lays particular stress on the latter point.
In those cases however where one is led to suspect that ulceration may be present in the colon, massage is of course contra-indicated. Escherich advises the wearing of an elastic abdominal binder as an addition to the treatment by massage.

Electricity.

This has been frequently used, generally along with massage, and apparently with benefit.

Lennander who especially recommends it, inserts one electrad per anum into the dilated colon and applies the other with a stroking movement to the abdominal wall over the colon.

Treatment of an Acute Obstructive attack.

Whatever may have been the cause of the disease to start with in a particular case, one may be fairly certain if it has one of these acute attacks that a certain amount of kinking is present, and that the repeated efforts of the bowel to overcome the kink are simply tending to exaggerate the obstruction. Short of surgical interference two methods of treatment are open to us.

(1) To overcome the kink by passing a long tube per rectum. A fairly rigid rubber stomach tube this should be tried in the first place but if will not pass/
pass the kink, a gum-elastic instrument should be tried. In numerous cases the passage of such a tube has allowed the sudden escape of a large quantity of flatus with great relief.

(2) To administer drugs which will lessen the intestinal spasm always present in such attacks.

Opium in some form is the drug of most value. Passage of flatus has frequently followed the relaxation caused by an opiate in cases during such attacks.

These two methods may very well be combined. Kredal suggests giving opium per rectum in acute attacks of ileus and also inflating the rectum with air to overcome the kink from below. I can find no record of the successful application of the latter suggestion.

Should medical treatment be unsuccessful in palliating the condition of chronic constipation with distension and toxaemia;

Should attacks of ileus recur frequently, or should any one of these attacks resist medical treatment, then surgical interference is indicated.

Surgical Treatment.

The/
Surgical Treatment.

The different operative methods which have been employed in cases of this disease are very numerous. They may be roughly divided into two groups.

I. **Radical operations** removing in whole or in part the offending portion of bowel, (amputation, resection) or reducing it in size (coloplication).

II. **Palliative operations** where the dilated bowel is left but where by drainage of it (colostomy), by fixation of it (colopexy), or by intestinal anastomosis with it, the operator endeavours to place the dilated bowel under such conditions that regular evacuation is secured and obstructive attacks avoided.

In the first place it will perhaps be instructive to review the different methods which have been employed, taking them up according to their severity in ascending succession.

Ito and Soyesima have made a fairly exhaustive review of this part of the subject and my figures are for the most part taken from their paper.

<table>
<thead>
<tr>
<th>Operation</th>
<th>No. of Cases</th>
</tr>
</thead>
<tbody>
<tr>
<td>I. Intestinal puncture.</td>
<td>10 cases</td>
</tr>
<tr>
<td>II. <strong>Exploratory laparotomy</strong> (nothing done or intestine) emptied per anum by compression)</td>
<td>20 &quot;</td>
</tr>
</tbody>
</table>
### III. Colotomy (evacuation of contents and then closure)
7 cases

### IV. Colostomy.
23 cases

### V. Coloplication (making folds in the long axis of the gut)
4 cases

### VI. Coloploste (Perthes in a case where a kink was present incised in long axis of bowel and stitched up in transverse axis as in Pyloroplasty)
1 case

### VII. Colopexy
6 successful, 3 unsuccessful.

### VIII. Enterocanostomosis
11 cases
- 6 successful
- 2 died from peritonitis
- 3 in status quo.

### IX. Resection.
13 successful, 5 died
- 1 in status quo
- 8 died some months later from diarrhoea.

### X. Amputation. (Treves)
1 case successful

Of these methods the ones which merit fuller consideration are Colostomy, Colopexy, Coloplication, Enterocanostomosis and Resection.

Resection is unquestionably the operation of choice and the best results have been obtained in those cases where it has been successfully carried out.

This operation is however frequently rendered very difficult if not impossible by the dilatation involving/
involving the lower part of the pelvic colon. In such a case one cannot get below the dilated portion but must cut through it and if one divides the bowel above on the proximal side of the dilated part, then there is great disparity in the size of the two cut ends to be united. This difficulty can be got over either by closing both the cut ends and then doing a lateral anastomosis or by partially closing the large lower end till it fits the other one, and then doing an end to end anastomosis just as was done in a Billroth's partial gastrectomy (the latter method was employed in Case V. of my series).

It were probably better however to restrict the operation of resection to those cases where the dilated and hypertrophied portion of bowel can be removed in its entirety. A case was recently reported where the whole colon was resected, the ileum being united to the rectum.

Short of resection what is the operation of choice?

The answer to this question cannot be a dogmatic one but is dependent in each individual case on the pathological anatomy of the colon.

Colopexy.
Colopexy.

If the dilatation and hypertrophy have not reached an advanced stage and especially if from the symptoms before operation and from the condition of parts found at operation one recognizes that kinking in the lower part of the pelvic colon had been a potent factor in causing the condition, then colopexy might with advantage be performed.

The lower part of the pelvic colon is pulled up and then fixed to the abdominal wall. The fixation of the colon must be very thoroughly carried out, otherwise the powerful hypertrophied bowel by its forcible peristalsis tends to break loose again.

Kredel recommends anchoring the pelvic meso-colon as well as the pelvic colon by stitches.

This operation is obviously only of value in a small proportion of cases.

Coloplication

This operation is indicated if the dilatation of the colon be found to be much in excess of the hypertrophy. This operation was eminently successful in a case of this type reported by Brooks.

Longitudinal tucks are made in the colon till it is reduced to normal or nearly normal size.

Cases suited for this operation are undoubtedly rare.

Entero-anastomosis /
Entero-anastomosis is indicated if the enlargement of the colon be not extreme and there be no history in the case of recurring attacks of ileus. Under the heading entero-anastomosis I refer particularly to the operation where the lower part of the ileum is anastomosed laterally with the pelvic colon.

Lateral anastomosis of the two limbs of a dilated loop of pelvic colon, and also of the pelvic colon with the rectum have been practised and a history of ileus would of course be an indication for one or other of these operations, but the first of them has not given good results and the second though sound in theory is extremely difficult in execution, so that I will not consider them further.

The principle of the "Ileo-colic" anastomosis is that the influx to the pelvic colon of the liquid content of the small intestine prevents the stagnation and gaseous distension which are the chief factors in producing the troublesome symptoms of this disease. The only points about the operation requiring mention are that the anastomosis be made large enough (at least 2" long) and that it be iso peristaltic.

In one case of this kind however a successful result/
result was only obtained after a second operation at which the ileum was divided distal to the anastomosis and the two cut ends closed.

Entero-anastomosis was successful in relieving Case III. of my series of all his symptoms. Colostomy.

This is indicated in many cases as a temporary measure when operation is performed for acute obstructive symptoms, and the patient's general condition will not permit of any more serious interference. Some writers, particularly Kredel, regard colostomy in this disease simply as an emergency operation and "would never recommend it in an uncomplicated case or as a permanent method of treatment."

I must however take exception to this statement as there is one type of case in which colostomy appears to me to be the only reasonable operation.

This type is something as follows: -

The case is a long standing one with great abdominal enlargement and suffering from recurrent obstructive attacks. On opening the abdomen an enormous pelvic colon is found with thick leathery hypertrophied walls. The dilatation involves the lower part of the pelvic/
pelvic colon so that one cannot get below it to resect, and its relation to the rectum is such that kinking in this region is liable to occur.

Nothing else than a small colostomy opening will ensure for such a case relief.

The colostomy opening though securely made need only be very small, as its function is merely that of a safety valve and not that of an artificial anus, that is to say that while it will relieve all flatulent distension by allowing gas to escape, the bowels will act by the normal channel.

Case V. of my series illustrates how successful this method of treatment may be.

The disagreeable state of having a permanent abdominal opening is more than compensated for in these cases by the freedom from chronic toxaemia and from obstructive attacks, which they enjoy.
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