HIRCHSPRUNG'S DISEASE ("IDIOPATHIC" DILATATION OF THE COLON).

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The name idiopathic dilatation of the colon, originally given by Gee to the condition which is discussed 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 of the subject it early becomes evident that the term "idiopathic" conveys to different minds different ideas, so that from the outset one has to work within ill-defined limits.

The Italian synonym "megacolon congenito" is more definite, but it implies more than is justified by our present knowledge of the disease. Probably the German name, Hirschsprung's disease, is as good as any, for it is frankly non-committal as to etiology, in regard to which certainty is as yet impossible.

By Hirschsprung'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 condition is recognised, and the dilated coil of bowel frequently appears to fill almost the whole abdominal cavity.

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 that of a man over sixty years of age.

Pronounced constipation, dating from birth or shortly after birth, characterises the condition, and abdominal distension and enlargement are marked features. The majority of the recognised cases have died in early childhood from intestinal toxæmia or from 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 malady 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 recognised. 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 120.

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

Since Hirschsprung in 1888 published his first two cases of this malady, and for the first time gave a clear and precise clinical picture of the disease, quite a considerable literature has accumulated on the subject, and very numerous and varied theories have been brought forward as to its etiology. Such diversity of opinion exists among competent observers that one is almost driven to the conclusion that the dilatation and hypertrophy of the colon which we associate with the name of Hirschsprung may be caused by any one of several factors, and that no one cause will be found which will account for all the cases reported.

One may here indicate briefly the various views which are held in regard to its etiology, and at a later stage enter more fully into this part of the subject.

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 Neugebauer all support this view.

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."

3. That an abnormally long pelvic meso-colon with 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.
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.

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, and Lunn, Fitz and Lennander also favour it.

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 the sphincter ani” to be the cause.

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

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 form.

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).

Morbid Anatomy.—The condition of parts is best studied from below upwards. The anus is normal, and the rectum is, as a rule, normal. In some cases the rectum has been found very roomy and dilated. Hawkins believes that the dilated colon always ends by a funnel-shaped narrowing, and that this funnel-shaped part is in many cases the upper half of the rectum. Lately, however, several observers have noted that below the greatly enlarged pelvic colon the rectum was found in a contracted state, suggesting muscular spasm.

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 it has not been found either before or since his publication.

Generally the dilatation begins just above the rectum, but the lower 2 to 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. As a rule, the pelvic colon is found enormously enlarged, having the form of a single loop extending upwards into the abdomen and also slightly to the right—in many cases extending up to 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 opening the abdomen it conceals all the viscera (with the exception of the liver) from view. It may have a diameter of from 6 to 8 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 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 portions of the colon were reached, but in quite a considerable number the descending colon has been dilated.

Less frequently has the transverse colon been found enlarged, and still less frequently the ascending colon and cæcum. 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.

In long-standing cases the bowel has a tough, leathery consistence, and in these cases there is found considerable fibrous hyperplasia in its wall, and evidence of "chronic interstitial colitis."

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-Geschwüre" (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 it is gaseous, with scanty semi-solid faeces; in a fair proportion, 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.—If there be no ulceration present, the mucous membrane shows only some degree of leucocyte infiltration, indicating a state of chronic irritation. The muscularis mucosae is usually slightly hypertrophied. Some leucocyte infiltration of this layer is the rule, and interstitial fibrous changes are frequently seen. The submucous coat always shows a much denser and more fibrous structure than normal, its blood-vessels are more numerous, and round them lymphocytes are collected in great numbers.

The circular muscular coat always shows an extreme degree of hypertrophy, being frequently four to five times its normal thickness. This increase in thickness is due to a true muscular hypertrophy, the muscle fibres being larger and more numerous than normal.

Leucocyte infiltration of this coat is generally seen, and its blood-vessels are increased both in size and number. In old-standing cases the muscle may be replaced to some extent by dense connective tissue fibres.

The longitudinal muscular coat generally shows some degree of hypertrophy, but is rarely more than twice its normal thickness.

The serous coat invariably shows some degree of fibrous thickening.

Symptoms and Signs.—The clinical picture presented by the disease is a 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 course, compensation being quite inadequate.

Others again are so well compensated that they present few 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. These shall be considered first.

Constipation is characteristically present, and almost invariably dates from birth. Generally it is of a pronounced type, and it is not uncommon to get a history that the bowels have not acted for several weeks. In many recorded cases there 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 medicines and injections, and in many cases never having a natural motion. When the child is weaned 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 scybalous, but, as a rule, are small, soft and semi-solid, and frequently very offensive. In some cases, however, a great quantity of hard scybalous faeces may collect in the colon, and small hard masses may be passed from time to time.

Flatulence and abdominal distension are always present to some degree. The distension of the abdomen is generally noted shortly 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 four other cases—those of Hobbs and de Richemonde, Mya, Futterer and Middeldorpf, and Schmidt—there was a history of abdominal enlargement at birth.

Duval collected twenty-nine cases where the time of appearance of the abdominal distension was noted—

| Period              | Cases |
|---------------------|-------|
| In the first few days of life | 12    |
| In the first month   | 2     |
| In the first six months | 6   |
| In the first year    | 4     |
| In the second year   | 1     |
| In infancy           | 4     |

29 cases.
The degree of the distension may be enormous, and may seriously interfere with respiration. In practically every case 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 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.

In a case running an acute course, 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 look, and is often drowsy and listless from toxæmia. 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 toxæmia, or from the perforation of an ulcer. In some cases acute colitis supervenes, with profuse diarrhoea and a rapidly fatal issue.

In the chronic type of case one finds obstinate 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 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.

In compensated cases 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 forty or fifty 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, 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. There is usually 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 into the right hypochondriac region. In 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 entirely gaseous; in rarer cases it may be dull, however, as in them the bowel is distended with solid fecal 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. 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 latter 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 hydronephrosis, from pressure on the left ureter, was present.

The prognosis varies 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.

Concetti collected thirty cases with a view to finding out the prognosis. In nine of these cases the result of treatment was unknown, as they were altogether lost sight of. Of the remaining twenty-one cases, nineteen died, two lived. Thirteen of the fatal cases died within the first three years of life.

It is thus evident that the time of greatest danger is the first few years of life, and that if a case can be 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 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 cæcum and the drawing in of the pelvic colon by the shortening of its mesentery.

There can be no doubt 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. Of thirty-seven cases which one has been able to collect from recent literature, a fatal issue ensued in nineteen. This shows a
death-rate of just over 51 per cent., a much more hopeful outlook than that presented by the older statistics.

Cases.

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

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., at 5 weeks. Admitted 7th July 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, 7 lbs. 10 ozs. Small pinched face. Nutrition poor. Abdomen is much distended; the skin tense and glazed, and 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 and hyper-
trophied, 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 broncho-pneumonia.

CASE II.—Lizzie N., æt. 2 years and 4 months. Admitted 3rd November 1900. 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 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 bowel for twelve inches, and the colon irrigated. Two copious evacuations resulted.

On 6th November 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 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 B., æt. 5½ years. Admitted to hospital on 23rd June 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. 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 constipation 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 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 anæmic, 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 tympanic note. No dulness in flanks. No thrill.
Per Rectum.—No spasm of sphincter. Rectum capacious, full of semi-fluid dark greenish faces. Nothing abnormal felt.

On 24th June 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 fine 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.

Four days later the bowel was opened and the mucous edges stitched to the skin. On opening the gut an enormous quantity of flatus escaped, followed by a considerable quantity of fluid faces, the abdomen flattening out considerably. For some days after this the child was very weak, and lay in a curious listless and apparently toxæmic 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 action regularly through the colostomy opening.

In February 1903 child readmitted, 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 construction of any sort to be made out, but only an enormously 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 food. Abdomen still abnormally large. (Fig. 2.)

Case IV.—William M., æt. 6 years. Admitted to hospital on 24th March 1906. Complaint—Constipation from birth. Swelling of the abdomen for the past twelve months.

History.—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 infracostal plane . . 23 inches.
" " umbilicus . . 22 "
Every now and then in either flank and across the upper part of the abdomen a swelling two to three inches broad stands out for a few moments and then fades 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 24th April 1906
Circumference at infracostal plane . . $24\frac{3}{4}$ inches.
" " umbilicus . . $23\frac{1}{2}$ "
Surgical interference was decided on, and on 10th May 1906 the abdomen was opened in the mid-line below the umbilicus. A greatly distended pelvic colon, very thick walled and fully five inches in diameter appeared, and was with difficulty delivered through the wound. The large intestine was examined throughout its whole extent. The cæcum was of practically normal 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 (eight to ten 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 $2\frac{1}{2}$ 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 the child was discharged six weeks after the operation. Bowels moved naturally once in four to five days, the motions being pale and semi-solid. An occasional enema still required. Remained in good health till 2nd August 1906, when he was readmitted with symptoms of obstruction, vomiting and distended abdomen.

On admission
Circumference at infracostal plane . . 27 inches.
" " umbilicus . . $26\frac{1}{4}$ "
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 tendency to obstructive attacks or distension.
FIG. 1.

Case III.—Before the bowel was opened.

Fig. 2.

Case III.—Five years after operation.
Hirschsprung's Disease

Case V.—James D., æt. 60 years. Admitted to hospital on 2nd May 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 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’s 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 the superficial veins are prominent. Tympanic 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, 2nd May.—An incision 3½ inches long made half an 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 trocar and cannula 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 cannula was now enlarged and a rubber tube was inserted and stitched to the bowel by two silkworm gut sutures. Skin incision now closed with silkworm 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 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, 15th May 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 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 obstructive 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 15th June patient was discharged, the bowels acting partly per rectum but chiefly through the colotomy opening in the right iliac region. After returning home 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 3rd January 1907 he again noticed that his abdomen was becoming distended, and obstinate constipation set in. On 4th January he again began to suffer from cramping pains in the abdomen, he passed no flatus and became more distended. On 5th January 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 6th January patient was again admitted to Chalmers’s 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-note is tympanitic all over. Every few minutes patient has a spasm of pain, when patterning of the abdomen becomes very well marked. A large
coils about four to five 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 twenty to thirty 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 January 1907.—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 five to six inches in diameter.

The abdominal cavity having been shut off with sterile swabs, the bowel was punctured with a trocar and cannula. 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 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 did very well. The stitches were removed on the tenth 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. Numerous drugs were tried for the chronic flatulence and spasmodic pain. Opium and calomel given together, and later β-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 28 cm. 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 gripping pains. Repeated enemata were of no avail, and on 3rd April 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. 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 trocar and cannula 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 cannula was left in situ, being fixed with strapping. On 5th April the spasms had returned and patient showed signs of peritonitis, therefore operation agreed on.

Operation, 5th April.—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 the old adhesions in this region had been stretched, and the colon was not in close apposition with the abdominal cicatrix. The cannula 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 trocar and cannula was now plunged into the bowel, and a large quantity of flatus escaped. Cannula 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 mucous membrane stitched to the skin edges of the wound so as to give patient a permanent 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 fecal 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 interstitial 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 forty 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.

Treatment.—It is generally recognised that to cure this disease is in the great majority of cases impossible. Our aim, therefore, must be to decide which are the most successful methods of palliative treatment.

Most observers are agreed that medical treatment alone is insufficient, and some form of surgical interference is called
for. However, though no case of actual cure of the condition by medical treatment has been recorded, several cases have shown very marked improvement under suitable medical measures. 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 recognised in its early stage when treatment of a purely medical nature is of undoubted value. On the other hand, the only completely successful results so far recorded have been obtained by surgical treatment. Medical treatment must always have two main objectives—firstly, to improve the muscular tone of the colon, and, secondly, to prevent stagnation and fermentation in the colon.

The question of the most suitable diet for such cases is not yet decided. Most writers recommend a diet which will leave as little residue as possible. It is, however, not unlikely that the giant colon could contract upon and propel onwards a bulky residue better than it could a small one.

In infants on the breast, weaning should be delayed as long as possible, as a change to artificial feeding almost invariably aggravates the condition.

Ordinary purgative medicines are of little value, but drugs which increase the muscular tone of the bowel, e.g. strychnine and belladonna, have been found to be useful. Cheadle has laid special emphasis on the value of drugs increasing intestinal tonus in cases of pronounced constipation in children, and many of the cases reported by him were probably examples of the milder degrees of Hirschsprung's disease.

Intestinal antiseptics or antifermentative drugs, such as β-napthol or sodium sulpho-carbolate, may be used with benefit. Unquestionably the medical measure of most value is lavage of the colon. This should be carried out at least once daily, a long rubber tube being passed per rectum right up into the dilated colon, and the latter irrigated with several pints of water. By this means fermentation and distension are to a great extent obviated.

Abdominal massage is of undoubted 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. Escherich lays particular stress on the latter point, and advises also the wearing of an elastic
abdominal binder. Massage is, of course, contra-indicated in all cases where one is led to suspect that ulceration is present. Electrical treatment has been frequently employed, generally along with massage, and apparently with benefit. Lennander, who especially recommended it, inserted one electrode per anum into the dilated colon, and applied the other with a stroking movement to the abdominal wall. Should an acute obstructive attack supervene, one may feel certain that whatever the primary cause of the malady may have been, there is now some kinking of the distended loop over the normal bowel below, and that the repeated efforts of the bowel to overcome the kink are simply tending to aggravate the obstruction. Short of surgical interference, two methods of treatment are open to us in such a contingency. One may pass per rectum a fairly rigid rubber stomach tube, and the instrument rounding the kink allows of the sudden escape of a large quantity of flatus with great relief to the patient. Secondly, one may 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 during such attacks.

These two methods may be very profitably combined.

If medical treatment does not succeed in palliating the condition of chronic constipation with distension and toxaemia, if attacks of ileus recur frequently, or if any one of these attacks resists medical treatment, then surgical interference is indicated.

Very many methods of surgical treatment have been practised, and much has been written on the subject. One need therefore only consider here the methods which are most frequently applicable and which have met with most success. These are—resection of the affected portion of colon, entero-anastomosis, coloplication, colopexy, and colostomy.

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 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 larger 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.—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 recognises that kinking in the lower part of the pelvic colon has 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, by stitches, the pelvic meso-colon as well as the pelvic colon.

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

Coloplication 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 is indicated if the enlargement of the colon be not extreme and there be no history of recurring attacks of ileus. Under the heading entero-anastomosis one refers 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 one need not consider them further.

The principle of the "ileo-colic" anastomosis is that the influx to the pelvic colon of the liquid contents 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 two inches long), and that it be iso-peristaltic.

In one case of this kind, however, a successful 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 the symptoms. Colostomy 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."

One must, however, take exception to this statement, as there is one type of case in which colostomy appears 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 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 toxæmia and from obstructive attacks which they enjoy.

Etiology.—The greatest interest unquestionably centres round the question as to what is the primary cause of the condition. Here we are at once beset with the difficulty that we do not at present know the pathology of the early stages of the malady, for by the time that cases come under medical supervision it is more than probable that secondary developments have obscured the primary cause.

To fully understand the views brought forward in regard to the etiology, one must bear in mind that in the new-born infant the pelvic colon and its mesentery are relatively long and lax. Thus, whereas 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 new-born child the similar relation is 1 to 1.7, and during the first two years of life the relation averages roughly 1 to 2. Moreover, in the infant the pelvic meso-colon has a comparatively long and extensive attachment to the posterior abdominal wall. These factors combine to give this portion of the colon a very wide range of mobility, and consequently one may find it occupying almost any region of the abdomen.

The question as to whether some cases are of nervous origin must remain for the time being sub judice, as no definite evidence for or against this view is available.

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 large colon with thick hypertrophied walls in any of these cases at birth, as Hirschsprung and Kredel believe. The only case where the signs and symptoms of this disease were present from birth is recorded by 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 three and a half 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. Escherich 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. In no case where a child has died during the first few days of life has the typical hypertrophy of
PLATE II.

Fig. 3. (After Perthes.)
Mechanism of kinking of pelvic colon over rectum with valve formation.

Fig. 4.
Abdomen of new-born child. Small intestines removed; water injected into the lower cut end of ileum; distension of colon most marked in its pelvic portion which fills the lower half of the abdomen.
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the bowel wall been demonstrated. (The case recently reported by Schmidt as one of truly congenital dilatation and hypertrophy is unconvincing.)

The anatomical theory held by many is, briefly, "that an exaggeration of the long sling-like pelvic colon, found normally in the infants, is the cause of the 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 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 photograph.)

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

Further, 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 diverti-
cula, found that true diverticula occur more frequently in the pelvic colon than in any other part of the intestinal canal. 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, injections through the fistula did not come away per anum—showing the presence 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 new-born 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, we must at once recognise that in a considerable proportion 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.

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 abdomens of newly-born infants one is struck by the fact that while the small intestine is usually more or less collapsed and empty, the large intestine, but more especially the pelvic colon, is almost invariably full of and is 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 kinking or rectal atresia was found.

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 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 over-distension has taken place at or before birth, one can readily imagine that this over-distended pelvic colon will not empty itself after birth so soon or so readily as normal, just as we find defective action in an over-distended bladder. Now we know that though meconium is sterile at birth, within twelve to twenty-four hours 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.

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. We may 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 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 there-
fore 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 from toxemia in infancy or early childhood. Where the hypertrophy is sufficient to compensate 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. In support of this, it is worthy of note that it is extremely rare to meet with a case 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 the 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 compensate for the dilatation, the patient may live for 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.

Bibliography.

A complete and up-to-date bibliography of the subject, compiled by Zesas, will be found in the Centralbl. f. d. Grenzgeb. d. Med. u. Chir., 8th and 22nd March 1909.
Fig. 5.
Pelvic colon of a new-born infant showing great distension with meconium.

Fig. 6.
Dilated pelvic colon in a child who died twelve hours after birth. The distended loop contained gas and meconium, fermentation of the bowel content having already commenced.