THE PHYSIOLOGICAL SPHINCTERS OF THE ALIMENTARY CANAL AND THEIR AFFECTIONS.

***
*

J. A. Munro Cameron.

ProQuest LLC.
789 East Eisenhower Parkway
P.O. Box 1346
Ann Arbor, MI 48106 – 1346

I. The Pharyngo-oesophageal Sphincter.

In the normal act of swallowing, it is customary to describe two stages: a voluntary stage in which the bolus lies in front of the pillars of the fauces, and an involuntary stage in which it passes onwards into the oesophagus. In the former, the contraction of the muscles of the lips and cheeks and the successive elevation of the tongue, hyoid bone, and thyroid cartilage, bring the bolus between the palatine arches. These are narrowed by the contraction of the glosso- and pharyngo-palatinus muscles, and the soft palate is tightened and brought in contact with the pharyngeal wall, so preventing an exit through the nasal cavity. The larynx is simultaneously closed by the action of the laryngeal muscles - arytenoid, thyro arytenoid and thyro-epiglottic - the arytenoid cartilages are approximated; the food then slips over the epiglottis and is gripped by the constrictors of the pharynx. The lowest of these, the inferior constrictor, and especially its lower half, the cricopharyngeus forms the boundary between the pharynx and oesophagus, and it is on affections of this area, the first of the physiological sphincters of the alimentary tract, that this study is based.

The/

The pharyngo-oesophageal sphincter is normally closed and only opens on deglutition; the closure of the muscle ordinarily prevents the swallowing of air. It is innervated by the pharyngeal plexus, - formed by the union of filaments from the glossopharyngeal, the vagus and the ascending pharyngeal branch of the cervical sympathetic - and branches are also supplied to it from the external laryngeal and recurrent nerves. A transverse section through the oesophagus just below their level shows that the nerves communicate directly with the muscles and that Auerbach's plexus is not interposed between the circular and longitudinal layers. This plexus appears, as far as my sections show, usually 1-2 centimetres below the entrance; Fig: 1 shows ganglia containing 3-5 cells, that is a common finding two centimetres down, and the number of cells in a ganglion increases until the cardia is reached, when 15-20 cells may be found in transverse and 30 in longitudinal sections. The epithelial surface is stratified and contains from 8-10 layers of cells, the nerve endings, some bulbous, some forked and some spiral shaped are uniformly disposed round the surface. The muscularis mucosae usually consists of two or at the most three layers of muscular tissue and in the submucous area, the tubulo racemose mucous glands and small collections of/

of lymphoid tissue are held together in a fairly vascular stroma which is strengthened by elastic fibres. The normal oesophageal glands are always external to the muscularis mucosae; sometimes however other glands are to be found which have the characters of the secreting mechanism in the cardiac end of the stomach and are therefore referred to as upper cardiac glands. In 10 out of 250 oesophagi examined I have found patches of these glands at the cricoid, varying from a few millimetres to nearly a centimetre in diameter, sometimes in one patch, occasionally in two symmetrical patches and rarely as a scattered collection of minute points.

The surface when the glands are aggregated in an area of any extent appears smooth and velvety, redder than and slightly sunk below the surrounding epithelium and may show at post mortem examination considerable auto-digestion.

Microscopically they appear as if fitted into the epithelium, the transition being always sharp, and they are limited externally by the muscularis mucosae. They contain oxyntic cells and presumably can produce acid.

The muscular zone consists of striated muscle and both longitudinal and circular layers are of similar width and do not run entirely in circular and longitudinal directions, many of the fibres having an oblique inclination. The longitudinal layer at the top of the oesophagus is

arranged/

arranged in three distinct bands, an anterior attached to the cricoid cartilage between the origins of the posterior crico-arytenoid muscles and two lateral, continuous with the fibres of the inferior constrictor. The anterior spreads out fanwise and soon blends with the fibres of the lateral bands, completely investing the tube.

It seems to me that on this anatomical continuity between the inferior constrictor and the longitudinal coat and not the circular coat, rests the production of peristalsis at the uppermost end of the oesophagus. Peristalsis anywhere in the oesophagus depends essentially on a relaxation of the area below the bolus and of contraction in the area above it, and normally is governed by the integrity of Auerbach's Plexus. In the situation under discussion where Auerbach's plexus is absent, the anterior band of longitudinal muscle acts as a brace to the oesophagus and the contraction of the inferior constrictor in its pull on the external bands, widens the area below it.

Affections of the Crico-Pharyngeus Muscle.

Any lesion which affects the oesophagus may be formed in this area, from the simplest inflammations to an acute diphtheritic process. It may be involved in the destructive/

destructive effects of corrosive poisons or impacted foreign bodies and may suffer from the presence of extra oesophageal inflammations or tumours. Its posterior wall, the thinnest part of the oesophagus, is the common site of pulsion diverticula, and any of these causes may give rise to dysphagia.

But it is with diseases of the sphincter that I am primarily concerned in this paper and before considering the most important of these - non-relaxation and spasmodic contraction - I shall just glance at two other affections of little practical importance:- paralysis and aerophagy.

Paralysis of the sphincter is always part of a general paralysis of the pharyngeal musculature and may be the result of a lesion in the medulla involving the centre for swallowing, or cortical when the muscular centres are involved by disease of the lower end of the ascending frontal convolutions or the knee of the capsule. In both, the lips and tongue are usually involved before the pharyngeal muscles, and while the lesion is usually inflammatory or embolic, paralysis may quickly follow the toxins of diphtheria or botulism. Once I have seen pharyngeal paralysis in a fatal case of cerebro-spinal meningitis and once, a convincing history of ptomaine poisoning upset by finding a small gumma in the floor of the fourth ventricle.

Short of fatal paralysis of this order there is a

modified paresis or lack of activity of the pharyngeal muscles and sphincter, which is not so uncommon and which produces the same effects as pharyngo-oesophageal spasm. This condition seems to consist of slight organic changes following possibly repeated pharyngeal inflammation and associated with a feeble reflex and feeble muscular response during the co-ordinated pharyngeal act of deglutition. The lesion might be compared with the paretic palate of chronic nasal catarrh where the palate is shrunken and almost immobile, the mucosa atrophic and the glands and musculature wasted. The reflex is diminished and the contractile power is feeble.

Aerophagy.

Air swallowing, as I have said, is normally prevented by the closure of the crico-pharyngeus, and if voluntary control of this muscle is learned, can take place at any time. The condition is more common as it is more important in veterinary than in medical practice. "Manger-biting", or "crib-biting", the term by which it is known in horses, consists in the animal biting repeatedly on the edge of the feeding trough and swallowing draught after draught of air until the belly becomes distended. The habit is usually of very little danger to the animal which sooner or later passes the air, but may be risky for the rider if the saddle girths be tied while distension is at its height.

In/

In human beings aerophagy belongs rather to the category of bad habits than disease. In two imbecile boys whom I had daily opportunity of watching, no ill effects of any kind ever seemed to be produced although one of them literally spent his day in swallowing and regurgitating air. At times it may be a cause of atonic dyspepsia and may even lead to cardiac distress or pseudo angina, but it seems to me that it generally is a habit which many children learn and with development as quickly forget, suffering no ill effects at all.

Non-Relaxation and Spasm of the Pharyngo-oesophageal sphincter.

These two conditions, differing only in the degree of obstruction offered to the transit of food, are separated from all other affections of this area by the characteristic anaemia and frequent enlargement of the spleen and liver, with which they are associated.

The two following cases are good illustrations:-

A married woman, aged 45, was admitted to the Victoria Infirmary, Glasgow, and stated that she had always enjoyed moderately good health until 5 years before, when she began to have difficulty in swallowing. At first there was inability to swallow pieces of meat and sometimes if she carelessly masticated crusts of bread a similar stoppage took place until she had washed the offending pieces over with/

with a draught of fluid. A week or two might pass in which there was no difficulty in deglutition but the condition gradually got worse until being afraid of choking at every meal, her diet became reduced to liquids with softened bread and butter. Two years after the first onset of dysphagia she became pale and was weak and easily fatigued, and became breathless on slight exertion.

On examination she appeared to be thin and the skin of her face showed a pallor not unlike that associated with pernicious anaemia. The lips were dry and pale and a fissure extended downwards from the junction of the lips on the right side. The mucous membrane of the mouth was pale and dry, and the tongue was smooth and devoid of papillae. The pharyngeal wall showed a similar waxy appearance and the lowest levels of the pharynx showed bands converging to the centre as if closing tightly the opening.

The heart sounds were soft but pure.

The spleen was palpated 3 finger breadths below the ribs and extended to within two inches of the umbilicus. The liver was just palpable below the costal margin.

The reflexes, deep and superficial, were found to be normal throughout and no alteration of pupils or of any of the special senses could be detected.

Blood/

Blood examination.

Red blood corpuscles, 2,500,000 per c.m.

White " " 5,000 " "

Haemoglobin . . . . . 15%.

Polymorphonuclear corpuscles . . . 50%

Lymphocytes 45%

Eosinophile . . . . . 2·5%

Hyaline . . . . . 2·5%

The red blood corpuscles stained evenly, were of normal size, showed slight poikilocytosis but no nucleated forms.

The Wassermann Reaction was negative and the Van den Bergh reaction was negative. No occult blood was detected in the faeces and the urine was free from any abnormal constituent.

Fractional Analysis of Stomach Contents.

	¼	½	1 hr.	1½.	2
Total acidity	10	15	25	35	40
Free HCl.	-	-	-	10	20
Starch	+	+	+	+	+
Bile	-	-	-	-	-

Blood was absent, mucin excessive. Microscopical examination revealed nothing of any importance.

Bacteriological examination of the throat showed streptococci and leptothrix filaments; blood cultures were negative. When she was discharged, recovered, streptococci were still present in the throat.

In/

In view of the strophic changes in the lining of the mouth and pharynx, the Hydrogen Ion Concentration of the saliva was estimated and PH was found to be 6-4, - slightly on the acid side of normal.

Treatment consisted in passing a bougie through the sphincter and the administration of a mixture of arsenic and iron. In three months the splenomegaly and enlargement of the liver had disappeared, she was eating without difficulty, meat, apples, prunes.and the red blood corpuscles numbered 4 millions with a haemoglobin percentage of 75.

Very little difficulty was encountered in passing the Einhorn tube so that this is probably an example of non-relaxation rather than spasm of the sphincter such as was found in the following:-

A single woman, aged 42, was admitted complaining of severe dysphagia and general weakness and debility. She was pale and ill looking and for six years had found that swallowing had become daily more difficult; at times she said she could swallow nothing. She attributed the onset of her symptoms to no particular cause and she had otherwise been in good health.

The red blood corpuscles numbered 3,600,000 per c.mm.

Haemoglobin 40%.

Leucocytes 4000.

Differential count, Polymorphs, 50%.
Lymphocytes 48%.
Eosinophiles 2%.

Wassermann Reaction - Negative.

A Fractional test meal analysis could not be employed as she could not swallow the Einhar tube.

Bacteriological examination revealed Streptococci in the throat swab, blood cultures were negative.

The spleen was enlarged and palpable but the enlargement was not excessive: the liver was just obvious below the costal margin.

There was general and rather striking pallor of the face and of the mucous membrane of the eyelids and lips. There was atrophic glossitis and the mouth generally was dry and parched.

P_H of saliva was 6-4.

Firm looking bands were seen in the parchment like pharyngeal wall, radiating to the oesophageal introitus.

No general anaesthetic was used for endoscopic examination and when the oesophagoscope entered the sphincter it was tightly gripped so that any further passage was prevented.

A second examination under chloroform anaesthesia was made and the sphincter stretched. After the subsidence of the pain and irritation of the mucosa consequent on the dilatation, she improved rapidly. Iron, Arsenic and small doses of Thyroid were administered and in a week or two no difficulty in swallowing food was encountered.

An/

with the closure of the pylorus by a well defined anatomical sphincter. Were there any real similarity between the closing of these two orifices one would expect that the administration of atropine would produce similar effects and that dilatation of the cardia would follow.

I have investigated this in a large number of normal subjects, in those who had diseases other than oesophageal, and in gastric and duodenal ulcers, in which the administration of atropin or belladonna before examination is a routine procedure of the radiologist. Every case examined was well under the influence of the drug whether judged by the dryness of the mouth or the state of the pupils.

Barium carbonate was administered in two strengths. In one, 2 oz. were suspended in a pint of water, so that a Barium drink resulted, the other was nearly of the consistence of porridge and about 5 oz. of Barium were generally used, although the actual amount differed from time to time without any substantial effect on the results, the essential point being that in one you had a fluid and in the other a semi solid medium. In swallowing fluids an unbroken wave traverses the oesophagus, halts slightly at the cardia and passes on to the anterior wall of the stomach, the whole process timed from the request to swallow/

An examination of the gastric contents revealed the presence of Free Hydrochloric acid, and a blood examination a month after she had left the hospital showed 4,500,000 red corpuscles, 6000 white. Haemoglobin 70% and the spleen was no longer enlarged.

These two cases reveal very clearly the limits of the pharyngeal condition encountered in this syndrome. The blood changes, the splenomegaly and slight enlargement of the liver are practically the same, but in one, the Einhar tube is passed relatively easily, the other resists the passage of an oesophagoscope. It is because of these variations in the condition of the sphincter that so much confusion has arisen in descriptions of this syndrome. The examples of non-relaxation were seen more often by physicians; the severe spasmodic cases by laryngologists. It is not therefore surprising that Plummer first called the condition hysterical dysphagia[1] and that Brown Kelly and D.R.Paterson[2] described it as spasmodic contraction of the pharyngo-oesophageal sphincter. At first sight we might easily imagine that the American and British observers were describing different conditions for Plummer and even more, Vinson,[3] stresses the anaemia and splenomegaly while the British laryngologists describe in detail the atrophic changes/

changes in the buccal and pharyngeal mucous membranes and the frequently unyielding nature of the sphincteric spasm although at the same time they draw attention to the pallor and anaemia, which they naturally conclude is secondary to the deprivation of food rich in iron and other essentials.

Age and Sex Incidence, and social condition.

In the present series of twenty-five cases all the patients were women, 15 married, 10 unmarried; the youngest was 41, the oldest 60; the average age was 47.1 years. (Table I.)

The condition is not apparently limited to any one class of the community. Some of the patients were poor and probably always undernourished; others were of good social standing and no fault could be found either with their surroundings or manner of living.

Syphilis and alcoholic indulgence can be excluded as causative factors and there appears to be no connection between this syndrome and the globus associated with hysteria. Many of the sufferers are nervous, worried looking, and easily upset by trifles, but they often state that their nervousness is due to fear of choking, and only commenced with the onset of dysphagia.

Symptoms.

Dysphagia is referred to the level of the larynx and the complaint is made that there is an unusual tightness of the throat or that something behind the larynx is gripping the food. Pain is not usually mentioned as in ulcerated conditions, and in only one case, No.22 of this series (Table I) was there referred pain. This patient stated that when the food was gripped she had also a pain in the back of the neck. The dysphagia is more exaggerated when the woman is tired and is more troublesome at an evening meal than at breakfast. In one case, whenever swallowing might become troublesome, she took a glass of water and if she regurgitated this some glairy mucus followed, deglutition then proceeded without any difficulty as far as that meal was concerned.

Sometimes there is a story of having been choked by a particular piece of meat, bread or vegetable, or of a piece of crust or a raisin having "stuck in the throat", but it is uncommon to hear that the throat has been scalded by hot food or fluid though sometimes dysphagia is dated from the impaction of a sharp substance such as fishbone.

At other times the onset of the disorder is gradual and sudden attacks are separated by intervals of varying length in which deglutition proceeds normally. The result, in the long run, is the same and food has to be masticated and swallowed with punctilious care; inevitably the/

the diet becomes reduced - meat, vegetables, fruit are banned; bread unless softened is avoided and tea or coffee and semi-solids become the daily fare. Even then there may be frequent catching of a fragment at the oesophageal entrance and distressing efforts are made to dislodge it, while regurgitation takes place if water is used to wash it down. The eating of meals amongst strangers becomes a nightmare, and conversation or any distraction from the immediate task of swallowing, is impossible.

In the milder and earlier cases, examination of the mouth and pharynx may reveal nothing beyond a pale and waxy looking mucous membrane with the tongue smooth and devoid of papillae. Later there are fissures extending outwards from the junctions of the lips, and the pharyngeal mucosa is parchment like, dry and atrophic with a tendency to bleed on manipulation. When the disease has lasted for some time, or when the symptoms are so acute that there may be difficulty in swallowing fluids, the endoscopic appearances are more definite. "It is found that the deepest part of the hypopharynx does not present the usual sphincter like appearance. Instead of rounded folds or cushions of mucous membrane forming a stellate arrangement, tense bands pass in/

in various directions with their thin edges pressed tightly together. The entrance to the oesophagus appears as a pinhole or small irregular opening and not in the middle line." (Brown Kelly).

At other times the opening appears as an obliquely placed slit, again it would seem to be closed by a web passing back from the cricoid. Dan Mackenzie (4) has reported two cases in which the lumen was reduced to a small circular orifice 3-5 m.m. in size, both of which resisted dilatation with the tube.

Vinson in his paper while agreeing that the mucosa of the mouth and pharynx is often dry and atrophic, states that no oesophageal web was ever seen by him and in this his colleagues Moersch and Conner concur, although the latter authors come nearer to the view of the British laryngologists when they remark that "while none of our patients complained of a lump in the throat, they said that during the periods when they were able to eat they could feel something relax in the throat. Might it not be possible that due to some nervous strain such a spasm occurs or is even brought on through an infection? In the final analysis, the condition may be found to be on the same basis as cardiospasm."

As regards the throat condition, it only remains to be said that one of the patients in the present series had also cardio spasm, presumably a reflex spasm because treatment of the condition at the upper end cured her, and

that/

that as a diagnostic point the dysphagia of cardio spasm is occasionally referred high up and the obstruction may be said to be at the laryngeal level.

Blood Changes.

With one exception, the pallor of these patients was striking and as distinctive as the facial appearance of pernicious anaemia; no degree of jaundice was ever seen. The hair in some cases was dry and brittle and in four cases there was seborrhoea of the scalp. The skin generally was moderately tense and no changes such as might be found in myxoedematous patients were ever encountered. These were specially looked for as the age of the sufferers and the original thyroid treatment of Plummer suggested that the thyroid secretion was at times deficient.

A summary of the detailed blood examinations is given in Table I and it will be seen that the changes there are essentially those of cholorosis or mild secondary anaemia with a constant and considerable reduction in the percentage of Haemoglobin. The red blood corpuscles are reduced but never to the same extent; the leucocytes with two exceptions, Cases 2 and 21, are well within normal limits and the differential count reveals nothing more than relative slight lymphocytosis.

Splenomegaly:

Enlargement of the spleen was found in eight patients but of the twenty five, seven were not examined. In my experience/

experience the enlargement is usually definite, the spleen being palpable on deep inspiration two or three inches below the margin of the ribs. In four cases the liver was also enlarged but never to the same extent.

Examination of the urine was negative and fractional analysis of the stomach contents, where it was possible to pass an Einhorn tube always revealed free Hydrochloric acid; microscopic examination of the stomach contents revealed no constant change, and occult blood was never found in the faeces.

Neurological examination was negative, the reflexes being easily elicited and though some of the patients were emotional and highly strung others might be described as almost lethargic.

Referred pain was only mentioned on one occasion and appeared to be quite definitely centred over the 5th and 6th cervical vertebrae.

The debility which is naturally associated with changes of this nature is seldom so outstanding as one finds for instance in prolonged starvation or continued toxaemia nor is the loss of weight very great. One patient had lost a stone in 5 years, another 8 lbs. in 3 years.

The signs of the syndrome may therefore be briefly summarised - glossitis and atrophy of the oral and pharyngeal mucosae, definite reduction in the haemoglobin percentage/

percentage, and some diminution of the red blood corpuscles, frequent splenomegaly and sometimes enlargement of the liver; and the symptoms - dysphagia referred to the level of the larynx, with loss of appetite, debility and nervousness.

Differential Diagnosis.

Although in practice this syndrome usually requires only to be differentiated from cancer of the hypopharynx, so far as throat conditions are concerned, consideration should embrace all forms of dysphagia at the upper end of the gullet - paralysis following bulbar or cortical lesions, ulceration from syphilitic, tuberculous or other infection, stricture following injury or the ingestion of a corrosive poison, cardio-spasm referred high up in the gullet, pulsion diverticula at the oesophageal entrance, aneurysm or mediastinal tumour, congenital webs of the oesophagus. In my experience, however, it is with none of these that confusion arises but with pernicious and secondary anaemia. Two of the twenty five examples recorded here were diagnosed as and treated for pernicious anaemia. In three others where secondary anaemia was the diagnosis, no complaint of dysphagia was made until long after treatment had been instituted. In one case the patient said that she didn't want to speak about her difficulty in swallowing, as she was afraid that an operation on her throat would be performed. Another was satisfied that for some years

past/

past, she had suffered from "a small swallow" and looked upon it as a dispensation of Providence. The third, to whom attention was drawn because of the long time she took over whatever food was placed before her, was told by someone that she had "rheumatics of the throat muscles" and from her experience of that ailment in the more mobile parts of her body was convinced that no amelioration was to be had from treatment.

The diagnosis is made on the negative results of Wassermann, X-ray and neurological examinations, the absence of clinical evidence of malignant disease, aneurysm or other obstruction to the onflowing oesophageal contents and the absence of any history of swallowing corrosive or other destructive liquid; while on the positive side there is anaemia without any substantial loss of weight, frequent enlargement of the spleen and liver, the presence of free HCl in the gastric contents, and the fact that the dysphagia is often as marked on the first day of the disease as on the last; finally the preponderance of these cases amongst women of middle life will naturally be considered.

As regards the value of X-ray examination, it is difficult to form a definite opinion. A lesion of the oesophagus or of the mediastinum will at once be apparent but I have found in watching these examinations of the pharyngo-oesophagus junction great difficulty in deciding what was actually

happening/

happening at the sphincter, and it is still more difficult to be certain of what factors were responsible for the delay occurring between the request to swallow and the appearance of the Barium in the gullet. In general terms these patients will not or cannot swallow the opaque meal, they equally refuse the Barium drink - one ounce of Barium to a pint of water, - if spasm of any degree is present. If non relaxation or perhaps slight abnormal tonus of the sphincter is present, delay between the request to swallow and the appearance in the oesophagus takes place, but one has no means of judging whether this is due to actual blockage at the sphincter or is the result of the dryness of the patient's mouth and a certain amount of repulsion to swallowing. The psychical effect of X-ray examination on many of these patients is in my experience harmful.

Treatment and Results.

The treatment is fortunately simple and effective and consists in passing a bougie or oesophagoscope or some form of dilator, according to the degree of resistance met with on examination. In very mild cases the passage of the Einhorn tube often makes swallowing easier for days afterwards. At the same time it seems necessary to impress upon the patients that benefit will result from the instrumental interference and should they be in hospital to see that

before/

before their discharge they do eat meat, vegetables, walnuts, prunes and crusts of bread. The passing of bougies blindly is generally deprecated in this country as false passages may easily be thus made in the submucosa. American authorities, however, do not lay such stress on obtaining a well illuminated view of the hypopharynx and state that the passage of any bougie is often sufficient. Here again, I think, the apparent contradiction is due to the different aspects of the syndrome which have been reported by the English and American authors. As a useful practical rule it seems to me that if the Einhorn tube passes relatively easily, treatment with any swallowed bougie will probably suffice, Hurst's weighted mercury bougie probably being as useful and as free from risk as any. If, however, the Einhorn tube cannot be passed, then the illumination of the hypopharynx should be as complete as possible and the dilatation performed by an expert and under general anaesthesia.

A mixture of iron and arsenic or iron alone is administered and usually as soon as the dysphagia disappears the haemoglobin percentage begins to increase and the spleen returns to its normal size. In a successful case this seldom takes more than 6-8 weeks. Plummer advised the administration of Thyroid extract for some time after the dysphagia ceased, on the ground that the prolonged starvation had resulted in a limitation of the body's need for/

for thyroid, and that a disuse atrophy of the gland had set in, and whether his theory is correct or not, the addition of small doses of Thyroid appears to hasten the disappearance of the pallor and anaemia.

None of the patients in this series has yet had any recurrence of symptoms, some have been well for more than two years. In this connection, though particularly dealing with serious and long-standing cases, Brown Kelly writes "At the first attempt to swallow after treatment, the patients felt as if something had been removed, and at once announced that they were cured. Two or three days later, when the soreness due to examination had passed off, they ate ham, prunes and other foods of which for years they had been unable to partake. They also found that they could converse at meals, and that they need not pay constant attention to mastication or be nervous about swallowing.

Some of the patients have had no return of their symptoms after an interval of one or two years; others after a few months have felt the need of care and have shown a slight tendency to choke. In exceptional cases, after an improvement lasting from one to three months, there has been relapse, but normal deglutition has been re-established - at least temporarily - by the passage of a bougie."

It is of interest in passing to note the effect of treatment by liver extract on this syndrome. Two of the/

of the cases, as I have said, were for a time treated as pernicious anaemia. In each case the number of red blood corpuscles increased rapidly: in one from 2,500,000 corpuscles to over 4 millions in 5 weeks: in the other from 3,000,000 to 4,400,000 in four weeks. In each case the haemoglobin percentage was practically unaltered, in the first Hb on admission was 15%, at the time of the second count 18%; in the second the Hb remained stationary at 40%. The pallor of both patients was unaltered; and they appeared to be more breathless on exertion as a result of treatment by this means.

Etiology and Pathology.

In any attempt to elucidate the pathology of this condition we are faced at the outset by the difficulty of determining whether the dysphagia precedes or is the result of the anaemia. In 15 of these cases dysphagia appeared to be the first symptom and at periods varying from two months to a year thereafter, the pallor was noticed by them or commented upon by their relatives. In two, it was difficult to ascertain the sequence of development, and of the remaining five, three thought that they had been pale before difficulty in swallowing developed and two were quite positive that pallor, unusual pallor and weakness preceded the dysphagia. Vinson, and

Moersch/

Moersch and Conner have had similar difficulty in investigating this point in America.

When anaemia precedes the dysphagia, we must presume that the atrophic changes in the oral and pharyngeal mucosae are analagous to the glossitis of pernicious anaemia and in some of the patients described here, the glossitis was that of pernicious anaemia. More often, I think, the dysphagia is the beginning of the disease and that the anaemia is the result of the difficulties of deglutition with the consequent curtailment of foods rich in iron and other essentials.

In a case of pellagra which I saw some years ago and which is not included in this series, dysphagia developed fairly early in the disease. It was not the result of any oral or pharyngeal ulceration, - not uncommon changes in pellagra - and was not, I now know, due to any paralysis of the pharyngeal muscles resulting from brain involvement, but was definitely the result of spasmodic or tonic contraction of the crico-pharyngeus arising in the course of the disease. It has been suggested by Hurst that a streptococcus in the throat is responsible alike for the anaemia and the dysphagia. Streptococci can often be isolated from the throats of these patients; so also can pneumococci; both are frequently enough found in any series of throats. But in four cases in which I have tried to agglutinate streptococci with the patient's serum, I have been/

been unsuccessful and the results of treatment would appear to negative this suggestion. Nearly all other observers agree that this syndrome results from a deficient dietary. We cannot, however, in any consideration of etiology, neglect the fact that practically every case occurs in women at or about the menopause, and the possibility of a vicious circle - a debility and anaemia associated with the climacteric, consequent changes in the pharyngeal mucosa, then dysphagia and limitation of food, with resulting accentuation of anaemia and splenomegaly.

The actual production of the dysphagia I take to be due to the effect of the atrophic processes on the nerve endings in the pharynx. These may be inflamed or they may be destroyed; the starting mechanism of the reflex arc is at fault and the swallowing movements suffer. At the same time the mucous glands in this region which normally facilitate swallowing are dry and may be atrophic; in short, the swallowing reflex is not normally and instantly initiated and completed. The block takes place at the cricopharyngeus and I have already mentioned that this muscle receives a branch from the superior cervical ganglion. How far this nervous supply may be responsible for the development of spasmodic contraction is an attractive but rather fruitless field of conjecture. In anxiety states

and/

and even ordinarily under the influence of fear or emotion, the difficulty of swallowing food is common knowledge and experience, and again the obstruction is quite definitely at the sphincter. The pathological basis of the form of dysphagia under discussion is in my opinion centred in the nerve endings and particularly the nerve endings on the mucosa immediately above the crico-pharyngeus. The first effect of their disorder is an achalasia of the muscle so that an interruption rather than a complete hold up of the swallowing movements occurs. Soon the unrelaxed muscle becomes subjected to direct pressure from the bolus and now probably spasm is elicited by this abnormal method of stimulation.

It is clear, if my explanation be correct, that the actual destruction of mucosa by the impaction of a foreign body, by ulceration or by malignant disease might produce spasm in exactly the same way and this, in fact, is found frequently.

Carcinoma of the Hypo-pharyngus.

No review of pharyngo-oesophageal spasm could be justly concluded without some reference to the frequency of carcinoma in this area in middle-aged women. From 70-80% of all cases of carcinoma of the hypopharynx are found in women, and it is frequently preceded for months or years by spasmodic dysphagia such as we have been discussing./

discussing. At other times the onset of the disease is more insidious and the first examination may reveal the ulcerated mass at the cricoid. The carcinoma is usually of the squamous celled variety although occasionally it may arise from the mucous glands. It is interesting, in passing, to note that no example arising from the upper cardiac glands has yet been described. The tendency of the growth is to encircle the oesophagus, and early to involve the lymphatic glands which drain this area - the deep cervical and pharyngeal.

It is difficult to believe that two conditions so definitely related in their age incidence and distribution are not even more intimately connected, and conceivably by a common cause.

There is no known difference in the histological appearance of the pharyngeal mucosa in the two sexes, and no greater liability to damage in women. So, it may be, that with the climacteric there is a proneness to changes in the pharyngeal lining which in one becomes manifested as a spasmodic dysphagia and in another leads to malignant proliferation.

REFERENCES.

***
*

(1). Vinson, P. - Minnesota Med. St. Paul, 1922, V. 107.

(2) Kelly, A.Brown. - Journ.Laryngol. and Otol. Edinburgh, 1919, xxxiv, 8.

(3) Paterson, D.R. - Ibid., Edin: 1919, xxxiv, 289.

(4) Mackenzie, Dan. - Ibid., Edin: 1918, xxxiii, 270.

(5) Hurst, A.F. - Guy's Host. Reps. 1926, October.

(6) Moersch, H.J. and Conner, H.M. - Arch: Otolar: Chicago, 1926, iv, 112.

(7) Cameron, J.A.M. - Quart. Journ. Medicine, 1928, Vol.xxii,85.

***
*

TABLE I.

Blood Examinations.

	Age.	R.B.Corpuscles. per c. mm.	White. per c. mm.	Hb.	Differential.
1	45	4,200,000	7,200	50	Normal
2	60	3,500,000	5,800	40	Leucopenia
3	50	3,800,000	6,600	50	Normal
4	55	5,000,000	7,200	60	Normal
5	45	3,850,000	6,400	50	"
6	47	4,000,000	6,200	55	"
7	50	3,820,000	6,100	55	"
8	52	4,160,000	6,400	58	"
9	45	3,840,000	6,800	60	"
10	50	4,200,000	7,000	55	"
11	46	4,400,000	6,800	52	"
12	48	3,600,000	5,800	48	Leucopenia
13	47	4,400,000	6,200	50	Normal
14	46	3,920,000	6,800	48	"
15	48	3,300,000	6,700	48	"
16	44	4,100,000	8,200	50	"
17	42	5,500,000	7,000	50	"
18	45	3,200,000	5,600	50	"
19	45	2,800,000	3,000	15	Leucopenia
20	42	3,000,100	4,000	40	"
21	50	3,500,000	6,000	50	Normal
22	44	3,400,000	5,000	40	Leucopenia
23	42	4,000,000	6,000	35	Normal
24	45	4,200,000	6,500	40	"
25	48	3,800,000	7,500	50	"
Average	47.6	3,972,000	6,270	47.6	

TABLE II.

General Examinations.

	Saliva P_H	Spleen.	Liver.	Free HCl	Throat Swab.
1	6.4	Normal	n.e.*	n.e.	Streptococcus & Leptothrix.
2	6.2	Enlarged	enlarged	n.e.	Streptococcus &
3	6.6	Normal	n.e.	n.e.	Streptococcus
4	6.9	n.e.	n.e.	n.e.	n.e.
5	6.4	Normal	n.e.	n.e.	Pneumococcus & Staph. Aureus.
6	6.6	Normal	n.e.	n.e.	Pneumococcus & Staph.Albus.
7	n.e.	n.e.	n.e.	n.e.	n.e.
8	6.4	Normal	Normal	Present.	Pneumococcus.
9	6.6	n.e.	n.e.	n.e.	n.e.
10	6.4	normal	Normal	Present.	n.e.
11	6.6	n.e.	n.e.	n.e.	Leptothrix filaments
12	6.4	Normal	Normal	n.e.	Pneumococcus & Staphylococcus.
13	n.e.	n.e.	n.e.	n.e.	n.e.
14	6.9	Enlarged	Normal	Present.	Streptococcus.
15	n.e.	Enlarged	n.e.	n.e.	Pneumococci & Staphylococcus.
16	n.e.	n.e.	n.e.	n.e.	n.e.
17	6.6.	Normal	Normal	Present	M.catarrhalis. Staphylococcus.
18	6.4	Enlarged	Enlarged	Present	n.e.
19	6.2	Enlarged	Enlarged	Present	Streptococcus, Leptothrix.
20	6.4	Enlarged	Enlarged	Present	Streptococcus
21	n.e.	Enlarged	Normal	Present	Pneumococcus, Staphylococcus.
22	6.2	Enlarged	Normal	Present	Streptococcus.
23	n.e.	n.e.	n.e.	n.e.	n.e.
24	6.2	Normal	Normal	n.e.	Pneumococcus. Staph.m.catarrhalis
25	6.4	Normal	Normal	Present	Streptococcus.

.*= Not examined.

2. THE CARDIAC SPHINCTER.

The increased interest manifested in disorders of the lower end of the oesophagus during the last few years has given currency to so many misleading statements regarding its elementary structure and function, that a re-investigation seemed desirable and appropriate. Authoritative statements, such as the following, made this the more necessary:-

> "The law of double innervation ruling in the vegetative system thus also applies to the oesophagus, on the one hand by the sympathetic, on the other by the parasympathetic (von.Greving)."

> "In the first place it is to be emphasized that the view of the antagonistic innervation of the internal organs by the parasympathetic and sympathetic systems, cannot be applied to the oesophagus (Hofer and Spiegel)."

Newly born children, and foetuses of six, seven and eight months have been used for dissection as ease of manipulation makes the following of nerves a comparatively light task, everything can be cut away that is not needed.

On the physiological side, no animal experiments have been performed as the work of earlier investigators has been generally confirmed, and is accepted by all.

X-ray examinations with varying concentrations of Barium carbonate have been made instead and a large number of normal and abnormal oesophagi have been screened.

In/

In the first place it is necessary to have a clear conception of the different areas of the lower end of the oesophagus. (Fig. 20)

The transition from the squamous epithelium of the oesophagus to the columnar epithelium is obvious to the naked eye, at the cardia, by the fact that a pale surface suddenly becomes redder in colour. From this line to the upper limit of the diaphragm is the cardiac canal. Above the diaphragm we distinguish the thoracic oesophagus, below the lower limit of the diaphragm, the abdominal oesophagus, between these we have the diaphragmatic oesophagus completely invested by the diaphragm on its posterior and lateral surfaces, surrounded but not to the same depth on its anterior aspect. The upper limit of this segment, the hiatal level, is the hiatal oesophagus. Just above the diaphragm the tips of the lungs lie close to the oesophageal wall and in deep inspiration must press upon it.

Nerve Supply.

The nervous mechanism of the upper limit of the oesophagus has been described in the previous study on the pharyngo-oesophageal sphincter. The cervical oesophagus is supplied by the recurrent laryngeal which usually enters the wall by a series of parallel branches. In the thoracic course the descending vagi join to form the oesophageal plexus, surrounding the gullet.

It is usually stated that a branch passes to this plexus from the great splanchnic nerve or ganglion. In none of my dissections have I found this connection, but there appears to be a fairly constant branch on both sides from the 4th sympathetic ganglion. Nor have I I found except once any branch going from the great splanchnic nerve to the oesophagus at all. In one example on the left side there appeared to be a short connection between the great splanchnic and the lower end of the oesophagus just above the diaphragm.

After the formation of the plexus the vagi resume their course, the left inclining to the front and the right behind and in this position enter the abdomen with the gullet, to supply respectively the anterior and posterior aspects of the stomach.

No direct branch from the sympathetic chain, great splanchnic or lesser splanchnic, from the level of the 4th ganglion to the diaphragm, was usually found, so that apart from any sympathetic branches which may be incorporated in the vagus from the cervical ganglia, or come indirectly to it through the recurrent, my dissections have failed to reveal any supply from the sympathetic system.

Below the diaphragm it is more difficult to be confident of the results of dissection as ganglia are so numerous and so closely associated, and the supply to the cardiac/

cardiac end of the stomach is considerable. But even to the abdominal oesophagus I could not follow twigs that were constant in each dissection and my conclusion is that the nerves from the sympathetic chain which reach this area, accompany the arteries, - the left inferior phrenic, and the left gastric - and may be subordinated only to their vaso-motor control.

The only remaining source of sympathetic supply is the phrenic which in addition to its origin from the 3rd, 4th and 5th cervical receives fibres from the cervical sympathetic chain, this root contrasting with the cerebrospinal fasciculus by its greyer colour. Hacques and Rousseaux in their investigation of the innervation of the lower sphincter of the oesophagus ascribe the sympathetic supply to this bundle. They distinguish a digestive diaphragm, essentially the muscle slings surrounding the oesophagus and a respiratory diaphragm, a term applied to the rest of the muscle. The former they state is involuntary and the latter voluntary. The sympathetic branch is continued to the phrenic plexus and from there the abdominal oesophagus receives its sympathetic supply. This hardly explains the sympathetic innervation of the diaphragmatic oesophagus. There is no continuity between the fibres of the diaphragm and the muscle of the oesophageal wall, on the contrary there is a distinct/

distinct separation by moderately firm fibrous tissue. Again the relations of the phrenic plexus on either side are slightly different; on the right side most of the fibres go to the inferior vena cava; on the left side they accompany the left inferior phrenic artery and with this vessel reach the oesophagus, but my impression is that they are confined to that vessel and do not innervate the wall.

My conclusion from my dissections is that there is no constant sympathetic supply to the oesophagus, from the upper limit of the stomach to the level of the 4th ganglion in the sympathetic chain. This view receives strong support from the experimental side. In experiments carried out by Williamina Abel in Glasgow it was found that section of one vagus nerve gave rise to distinct delay in the transmission of food through the oesophagus and to persistence in the tube of some of the swallowed contents for a considerable time. Double vagotomy, (one vagus being cut below and the other above the origin of the recurrent laryngeal nerve) led to complete stagnation of the Barium Meal with closure of the cardia, and in time to dilatation of the oesophagus.

Hofer and Spiegel confirmed these results and also showed that it was impossible to demonstrate any change in the peristaltic oesophageal movements in cats

or/

or rabbits, despite the bilateral removal of the superior cervical ganglia, cervical sympathetic, the stellate ganglion and splanchnic nerves. Further they showed that the changes produced by section or interference with the vagi developed whether or not the sympathetic nerves were retained.

Dilatation of the oesophagus very rapidly developed in rabbits but took longer in cats. In the latter after vagotomy, the lower half of the oesophagus becomes funnel-like from increased tonic contraction of the cardia but single peristaltic waves are observable, which drive small quantities into the stomach. Apparently the lower third of the oesophagus in the meat eating cat resists stretching for a longer period than the plant eating rabbit, as indeed might be expected from the thicker muscularis mucosa in the former animal. In dogs, too, spasmodic contraction of the cardia following vagotomy leads to a spindle shaped dilatation similar to what is seen clinically in man.

Histology.

A transverse section of the upper end of the oesophagus has already been described and examination of a section through the cardiac canal reveals no great differences apart from the essential ones, that in this area a collection of nerve cells - Auerbach's plexus - is present/

present between the circular and longitudinal muscle bundles and that the muscle fibres are unstriated. (Fig. 2)

The layers forming the epithelium are generally more numerous, the muscul/ous/ mucosa/ is somewhat thicker; there is rather more elastic tissue in the submucous layer and cardiac glands are to be found at times situated in it.

The muscular coats are of nearly equal thickness and the ganglia, often 7-9 in number, are spread equidistantly around the tube. If the view of a single ganglion is required sections made in an oblique rather than a transverse direction are the better procedure. No layer of nerve cells corresponding to Meissner's plexus in the submucosa of the intestines is found in the oesophagus. My preparations (Fig. 3) stained by Bi[illegible]chowsky method show that the nerve strands after coursing over the circular muscle bundle sometimes unite to form a thin layer in the submucosa before finally sending out the nerve endings to the epithelium, and occasionally there appears to be a nerve cell here and there but my impression is that these are rather swellings at the junctions of fibres or on the course of single fibres, than nucleated cells. They are certainly never seen in any sections except those stained with silver.

In/

In Auerbach's plexus, the nerve cells increase in number from the upper end of the oesophagus downwards.

In the topmost centimetre I have never seen nerve cells, seldom in the next centimetre and then often one cell in a ganglion. Two centimetres down they usually number 3-5 cells to the plexus and at the cardia may number between 20 and 30. In longitudinal sections the number is sometimes higher, occasionally as many as 40 cells being found, arranged in a sickle or half moon formation. These cells I have found in a foetus of five months but not in one of earlier development. By this time too, the permanent epithelium of the oesophagus has been formed.

It is interesting in passing to note that when cardiac glands are found in the oesophagus at either the cricoid or the cardia, they must have been formed by this time. According to Schridde the permanent epithelium makes its appearance in the seventeenth week of foetal life, "and in one embryo of 105-110 millimetres he found a group of five tall cylindrical goblet cells among the squamous basal cells in a recess at the side of the cricoid cartilage. They represent the first "anlage" of the cardiac glands. In the youngest embryos examined the oesophagus is lined by a single row of clear cubical endodermal epithelium. By about the fifth week (13 mm.) it

has/

has differentiated into two layers of clear cylindrical cells. During the tenth week (length 44 mm.) ciliated cells appear. In the twelfth week the lining consists of three or four rows of clear cubical cells, resembling those of a transitional epithelium. During the seventeenth week the cells of the basal layer become fibrillated in the manner of squamous epithelium; they divide and give rise to the permanent epithelium of the oesophagus. He insists strongly that it is only the cells of the basal layer that divide and give rise, step by step, to these different kinds of epithelium. The tall cylindrical clear cells, the ciliated cells and the clear cubical cells are in turn desquamated and lost." (Nicholson).

The muscular layers round the cardiac canal are usually exactly circular and longitudinal in direction very few oblique fibres being found and I have never seen any increase of thickness such as is to be found at the pylorus, except in cases of cardiospasm. Sometimes there is a distinct backward bend of the lower end of the oesophagus just before it enters the stomach, and at the same time there is a slight natural twistof the oesophagus upon itself. These factors may be of importance together with the amount of elastic tissue in the wall, in the normal closure of the cardia. A good deal of discussion has taken place as to whether the cardiac

canal/

cardiac canal is usually closed like the upper sphincter and only opens when peristalsis is in progress. Under X-ray examination it would certainly appear to be so, but when an oesophagoscope is introduced, the sphincter is then seen to be opening and closing and these movements are synchronous with respiration. The introduction of the tube and the stimulus to the oesophageal nerve endings may be responsible for this; in my opinion, from the findings in cases of cardiospasm, it is; on the other hand it may be due to the stretching of the walls involved in manipulation and the consequent straightening out of the deep pleats normally seen here in the mucosa, so that extra oesophageal factors are brought into play. The most obvious of these is the pressure of the tips of the lungs. That this part of the tube must be normally closed and opens ordinarily during peristalsis, under the pressure of fermentative or other gases from the stomach or during vomiting, seems the only reasonable means of explaining the delay in peristalsis at the cardia. Hyrtie described an incomplete anatomical sphincter and recently Hurst has figured a very definite one in a newly born child. These, however, must be of the greatest rarity and are probably due to local irritative conditions or to developmental phenomena. They should certainly not be used as the basis for a theory that the closure of the cardia is comparable

with/

with the closure of the pylorus by a well defined anatomical sphincter. Were there any real similarity between the closing of these two orifices one would expect that the administration of atropine would produce similar effects and that dilatation of the cardia would follow.

I have investigated this in a large number of normal subjects, in those who had diseases other than oesophageal, and in gastric and duodenal ulcers, in which the administration of atropin or belladonna before examination is a routine procedure of the radiologist. Every case examined was well under the influence of the drug whether judged by the dryness of the mouth or the state of the pupils.

Barium carbonate was administered in two strengths. In one, 2 oz. were suspended in a pint of water, so that a Barium drink resulted, the other was nearly of the consistence of porridge and about 5 oz. of Barium were generally used, although the actual amount differed from time to time without any substantial effect on the results, the essential point being that in one you had a fluid and in the other a semi solid medium. In swallowing fluids an unbroken wave traverses the oesophagus, halts slightly at the cardia and passes on to the anterior wall of the stomach, the whole process timed from the request to swallow/

swallow until the fluid passes the cardia being in the neighbourhood of 3 secs., the limits of my findings in normal people without atropine were from 2 $3/5$ - 5 seconds. In watching this descent one has the impression that no factor in the oesophageal wall is invoked at all and that either the pressure of the fluid alone dilates the canal in its transit, or that the initial momentum derived from the pharyngeal constrictors is sufficient.

When atropine was given no change was noted in the oesophageal wave, the time taken (in the few normals to whom atropine was given) was sometimes very slightly increased, but not any more increased than one sometime finds in successive examinations in the same subject.

In gastric and even more markedly sometimes in duodenal ulcers, the time was usually increased, the delay taking place at the cardia and irrespective of whether atropine had been administered.

If the presence of a gastric or duodenal ulcer were producing increased spasm or hypertonicity of the cardia atropin did not affect it.

In the swallowing of the semi-solid opaque medium the whole mechanism of oesophageal transmission seems to be different. Immediately after passing the crico-pharyngeus the bolus is pushed onwards in the oesophagus by a series of broken segmental waves, sometimes tarrying

as/

as if the load were too heavy for immediate transit and then passing more rapidly over one or two segments. At the cardia there is a pause; the lower end of the oesophagus is pulled down into a narrowed tube and suddenly gives way with the passage of the bolus into the stomach. One cannot but conclude that peristalsis with this consistence of Barium is dependent on successive stimulation of the nerve endings, that the reflex is carried to Auerbach's plexus and completed by the contraction of the circular muscle above the bolus and dilatation of the tube below it, presumably by the pull of the longitudinal coat.

The time occupied in transmission varies in normal people from about 7-10 seconds and of that at least 3-5 seconds are usually spent at the cardia.

After the administration of atropine the period which elapses in transmission is always considerably longer and the delay is general; it is not confined for instance to the cardia. There is increase in the time that elapses between the request to swallow and the appearance in the oesophagus; there is delay and more irregular contraction through the tube, and at the cardia the ordinary period of 3 or 4 seconds delay may be 6 or 8 or even longer, and if a gastric or duodenal ulcer be present, the delay may amount to two minutes. Unlike the pylorus then, the cardia is not relaxed by the administration/

administration of atropine.

The delay that has been noted in these observations is in my opinion not nervous in origin; it is certainly not entirely nervous, but is due in chief part to the drying up of the mucous glands in the pharynx and oesophagus. The essential differences manifested in the swallowing of fluids and solids or semi-solids, seems to me to be reducible to the amount of friction exercised in either case, on the oesophageal wall. A watery bolus with friction between it and the oesophageal epithelium at a minimum has the oesophagus almost passively opening as it travels. A semi solid or solid substance is exerting pressure on the wall throughout its progress, and evokes and necessitates individual contraction of successive segments to complete its transit. Friction is ordinarily reduced by the lubrication of the mucous glands and if these are inactivated by atropine the peristalsis of a Barium meal becomes a series of successive, jolting and almost discontinuous movements.

While the production of peristalsis by a reflex through Auerbach's plexus (the integrity of which is of course dependent on the vagus) is clear, the development of spasmodic contraction of the oesophageal muscle, as indeed of spasmodic contraction of any muscle, is far from clear.

Definite over contraction of the oesophageal muscle is found in true cases of cardiospasm, is usually associated with/

with carcinoma or ulceration, and generally with the impaction of a foreign body for any length of time. Any foreign body which has a smooth surface and therefore not likely to stick in the wall of the gullet will tend to come to rest at the particular point in which it is exerting greatest pressure upon the wall. Normally this would lead, were excitation of the nerve endings the only factor to be considered, to dilatation of the area below it and to contraction above. This no doubt at the actual moment of impaction is attempted, but the dilatation below is insufficient and the substance remains. Soon, spasmodic contraction develops and one can only conclude that this is due to (1) either unusual pressure on, or prolonged excitation of the nerve endings, or (2) the distension of the lumen of the gullet is permitting direct action on the muscular fibres.

Bayliss and Starling in their classical experiments on peristalsis found that when a repeated stimulus was given to any point in the intestinal wall, the distal relaxation (which followed a single stimulus) did not take place, contraction only followed. The implication of this experimental finding is that with all nervous and muscular processes intact, the area of relaxation below the bolus is the first part of the peristaltic wave to show impairment while the other part of the wave, the

contraction phase, remains unaltered. In this we may have a guide to the explanation of spasmodic contraction of visceral muscle and the factors concerned may be summarized thus:-

(1) Where there is a balanced dual nerve supply, overaction of one set of fibres may produce spasm, e.g. - overaction of the sympathetic by direct stimulation, or by relative inactivity of the vagus.

(2) Unusual pressure, or prolonged pressure on the nerve endings of the mucosa.

(3) Direct pressure on the muscular fibres, normal excitation being in abeyance, through pathological changes either in the ganglia, or in the nerve fibres or endings.

AFFECTIONS OF THE CARDIA.

Cardiospasm and Achalasia of the Cardia.

Cardiospasm was the name applied by the earlier investigators, Mihulicz, Zenker and von Ziemssen to a condition which led to dilatation of the oesophagus and was unattended by any distal obstruction, anatomical or pathological stenosis or stricture, and was therefore presumably due to spasmodic contraction of the cardiac sphincter. The effect of this contraction was to delay the passage of food into the stomach, and in time to dilate the walls of the gullet by the retention of unaccustomed contents. Another view, held by Morrell Mackenzie in this country, was, that there existed a myasthenia or at least a diminished contractile power of the oesophageal musculature. One early and unusual case described by Kraus revealed degeneration of the Vagus Nerves. In another examined by Klebs, changes of a degenerative character were found in the muscular coats and other authors suggested that the dilatation was due to developmental anomalies leading to a "trapping" or obstruction of contents at the cardiac sphincter, either by a V-shaped bend of the lower oesophagus itself or by pressure from an accessory liver/

liver lobe.

Extrinsic causes were also exploited and the most obvious of these contraction of the diaphragm - the pinchcock theory of Chevalier Jackson - held the field for some time until examples in which the dilatation extended well below the diaphragm, began to accumulate. A later school looked for endocrine disorders as an explanation and it was suggested that a lesion or defect or overaction of the suprarenal medulla altered the adrenalin supply and so caused spasm of this sphincter. In 1919 Brown Kelly and Irwin Moore had collected several oesophagi from fatal cases of this condition and some modification of previous views was obviously essential.

(1) The muscle of the oesophagus was in the majority of cases hypertrophied, and this increase was sometimes greatest at the maximum point of dilatation; it was never thinned there as might be expected in the dilatation of an asthenic muscle.

(2) Sometimes the muscle of the cardiac canal was hypertrophied; more often it was not.

(3) The dilatation sometimes extended through the abdominal oesophagus.

(4) No obstruction, developmental or otherwise, was found by them.

Shortly after this, Hurst revived the older view of Meltzer that there was not spasm of the cardiac sphincter. He/

He described a case of a newly born child with distinct increase of muscular tissue, (as I have already briefly noted) extending from the cardiac junction upwards for rather more than one inch, and stated that the so-called cardiac spasm of other writers was due to non-relaxation or achalasia of this sphincter. That the peristaltic wave in its descent was obstructed by the muscle not responding as it normally does by dilating. In none of the specimens examined by him was there overgrowth of the sphincter such as would be expected with spasmodic contraction and was, in fact, found in pylorospasm. He constructed a bougie weighted with 22 ounces of mercury, which always passed through the "obstruction" without difficulty. In 1926 Rake examining an oesophagus which was not dilated but considered to be an example of "compensated achalasia" described subacute inflammatory changes in the cells of Auerbach's Plexus.

Only one other point in the history of this condition need be mentioned. From the result of a large number of endoscopic observations Brown Kelly stated, that on introducing the tube in a normal subject, as it advanced towards the cardia, he found the orifice to be opening and closing with the movements of respiration. In cardiospasm, however, as soon as the oesophagoscope approached the cardia, tight closure of the muscle took place/

place and he naturally concluded that just above the cardia there was hyperaesthesia of the oesophageal wall.

This is a brief, but I think fairly complete statement of the position in regard to cardiospasm, until the work embodied in this thesis was undertaken. Microscopic examination of three excellent examples of cardiospasm with wide dilatation of the oesophagus was begun and the results were communicated by Brown Kelly in his Semon Lecture in 1926.

Since then six other fatal cases have been investigated, two by the courtesy of Dr Irwin Moore, and the original findings have been verified and expanded. These results have been confirmed by other observers.

The view submitted is, that cardiospasm and achalasia of the cardia, the two conditions only differing in the degree of tonicity of the muscle wall, are the result of interference with the nerve cells in the intermuscular plexus by inflammation or degeneration. That the lesion of the nerve cells is usually the result of an outward spread of inflammatory cells from a primary oesophagitis, but that sometimes the ganglia are involved by the specific selection of bacterial toxins paralleled by what is found in the paralyses which follow diphtheritic or meningococcal infection. These ganglionic changes are confined to the cardiac canal and are not found

elsewhere/

elsewhere in the oesophagus.

The immediate effect of the interference with the ganglia is failure of the muscular wall to respond to the stimulation of the nerve endings, alternatively the area of relaxation which should develop below the stimulated spot is in abeyance. As a result food sooner or later collects in the oesophagus and the wall of the viscus dilates, but in an attempt to expel the contents, the muscle wall and especially the circular layer of it hypertrophies. The hypertrophy indeed often appears to develop proportionately and pari passu with the dilatation. At times the sheer weight of the oesophageal column clears the obstruction and it wholly or partly passes into the stomach.

By unusual pressure on the nerve endings just above the cardia, assuming that they are not destroyed, no effect should be produced as the controlling cells are destroyed but direct pressure can be exerted on the muscular walls, and in this way I think spasm is produced. It is suggestive, in this connection, that in Brown Kelly's endoscopic observations, only when he pressed on the wall of the gullet above the cardia was tight closure produced. Higher in the tube where the ganglionic cells are apparently healthy, there is often visible, on X-ray examination, a characteristic rapid almost quivering

contraction/

contraction of the muscle as if it were contracting at its maximum power to press onwards the accumulated food. So rapid is this contraction that I have seen in a photograph, blurring of the wall.

It follows from what I have said that a lesion such as an ulcer or involvement of the mucosa and epithelium by a malignant process might interrupt the conduction of impulses from the nerve endings to the ganglia, and so might produce achalasia of the cardia, although the nerve cells might show no change. Abnormal stimulation of the nerve endings might similarly cause spasmodic contraction.

Before detailing the cases, the measurements from twenty normal oesophagi are tabulated:-

MEASUREMENTS OF OESOPHAGUS.

Taken from 20 adult cases without oesophageal disease.

	Age.	Thickness of wall.	Thickness of Muscular layers.	Diameter of cardia.	Length of Oesophagus.	Body length.
1	25	4 m.m.	2 m.m.	5 c.m.	24 c.m.	5 ft.3 ins.
2	27	3.8 m.m.	2 m.m.	4.5 c.m.	25 c.m.	5 ft. 8 ins.
3	29	4.2 m.m.	2.2. m.m.	5.5 c.m.	25 c.m.	5 ft.10 ins.
4	36	4.6 m.m.	2.4 m.m.	4.5 c.m.	23 c.m.	5 ft.
5	42	4 m.m.	2 m.m.	5 c.m.	27 c.m.	5 ft.10 ins.
6	18	3.8 m.m.	1.8 m.m.	5.2 c.m.	26 c.m.	5 ft.10 ins.
7	25	3.6 m.m.	1.8 m.m.	5 c.m.	24 c.m.	5 ft.2 ins.
8	46	3.8 m.m.	2 m.m.	4.5 c.m.	24 c.m.	5 ft.4 ins.
9	50	4.2 m.m.	2.2 m.m.	5.5 c.m.	25 c.m.	5 ft.6 ins.
10	42	4.4 m.m.	2.2 m.m.	6 c.m.	25 c.m.	5 ft.6 ins.
11	44	4.2 m.m.	2.2 m.m.	5 c.m.	25 c.m.	5 ft.8 ins.
12	42	4.6 m.m.	2.6 m.m.	5.5 c.m.	26 c.m.	5 ft.10 ins.
13	50	5 m.m.	2.5 m.m.	6 c.m.	25 c.m.	5 ft.6 ins.
14	64	5 m.m.	2.5 m.m.	6 c.m.	26 c.m.	5 ft.10 ins.
15	52	5 m.m.	3 m.m.	5 c.m.	25.5 c.m.	6 ft.
16	35	5.5 m.m.	3 m.m.	5 c.m.	24 c.m.	5 ft. 1 in.
17	37	5.5 m.m.	2.5 m.m.	5.5 c.m.	25 c.m.	5 ft. 4 ins.
18	39	4.5 m.m.	2.5 m.m.	6 c.m.	26 c.m.	5 ft. 5 ins.
19	41	5 m.m.	2.2 m.m.	5.5.c.m.	25 c.m.	5 ft. 6 ins.
20	43	5.5 m.m.	2.5 m.m.	4.5 c.m.	24 c.m.	5 ft. 1 ins.
Average		4.3 m.m.	2.3 m.m.	5.4 c.m.	25 c.m.	
Limits:		3.6 - 5.5 m.m.	1.8 - 3 m.m.	4.5 - 6 c.m.	24 - 27 c.m.	

Case I. J.K. female, aged 38, died from cerebral haemorrhage following a short illness. Her friends stated that she had had great difficulty in swallowing food for years and at times could take nothing at all.

At post mortem examination the oesophagus measured 25.5 c.m., and showed a diffuse spindle shaped dilatation, tapering to normal 5 c.m. below the cricoid level. 9 c.m. above the diaphragm was the site of maximum dilatation, the measurement there being 9 c.m. wide. At the cardia there was slight thickening, the muscle coat measuring 6 m.m. at its widest part the muscular wall measured 9.m.m. The thickness of the upper wall was 4 millimetres and the calibre of the cardia was 4 centimetres. On opening the oesophagus $\frac{3}{4}$ of a pint of fluid and food were present in the oesophagus. There were numerous small erosions of the mucosa especially in the lower half of the organ and the epithelium generally looked patchy and was easily peeled off.

Microscopic Examination.

Through Cardiac Canal.

The epithelium was only found in minute scattered patches. The submucous coat showed small ulcerated areas of various depths, and all the vessels were engorged while endothelial cells and lymphocytes were increased in their vicinity. The muscularis mucosa was increased

somewhat/

somewhat in thickness and fibrous changes in some of the bundles were noted. The circular muscle layer was manifestly hypertrophied and many bundles showed fatty and fibrous degeneration. The longitudinal coat showed similar changes but in less degree. Around the ganglia fibrous tissue had been deposited everywhere and in one part no ganglia at all were found. In those ganglia in which nerve cells were present degenerative and disintegrative changes had occurred and inflammatory cells were found lining the ganglionic capsule.

The vessels generally, as indeed throughout the body, were distinctly thickened but a Wasserman Reaction during life was negative.

2. Through wall at maximum dilatation.

The epithelium of the sections is again seldom seen and inflammatory cells in the submucous coat are frequent but do not penetrate very far. The muscular coats again show distinct hypertrophy but the ganglionic cells are not altered, staining quite well and being free from any inflammatory involvement. All the vessels in this area of the oesophagus show very definite sclerosis and the degenerative changes in the muscle fibres generally appear to be due to this cause.

(Figs. 4)

Case II. Oesophagus from a man of 21.

This patient had been admitted to hospital with dysphagia, which had begun suddenly, according to his statement, and apparently without cause, six years before. The obstruction was complained of opposite the lower end of the sternum and food reached this level without difficulty, but a great part of it was at once regurgitated. At meals, in order to get food down he had to drink freely, and on occasion, to stand, stretch or contort himself. During the ten days before admission the stoppage had become almost complete.

On inspection, the oesophagus was found to contain a good deal of fluid and food debris. Its diameter was increased and the hiatus appeared as a long narrow slit which opened and closed rhythmically so long as it was not touched by the tube but closed spasmodically when this was pushed against the wall. Firm and steady pressure failed to gain entrance to the stomach. Subsequent attempts to pass bougies through the cardia also failed.

In hospital, his food was measured and nearly all was regurgitated. Thus on one occasion he took 66 oz. and returned 59; on another he took 55½ oz. and only 3 were retained.

Gastrostomy was inadmissible and he died mainly from starvation.

Pathology./

Is this figure? as ref. at end of demo fig 5.

Pathology.

The oesophagus measured 19 c.m. from the cricoid to the ?tal level and presented a dilatation of fusiform type inclining slightly to the left. The wall was thickened in its whole length but especially at its lower end; above it measured 5 m.m. and in the last 7 or 8 c.m. 8 m.m. in thickness. The mucous membrane of the upper 1/3 was rugose and slightly thickened; in the lower 2/3 it was smoother and the mucosa was thickened and mamillated, the outgrowths firm in consistence varying from pinhead size to that of a small bean. Below the diaphragm the muscle was again thickened, attaining a maximum of 4.5 m.m. At a distance of 3 c/m. (below the diaphragm) the thickened muscle shaded into the muscular fibres of the stomach.

On inspection in situ, no band, adhesion, narrowing of the diaphragmatic opening or other condition likely to cause stenosis, was apparent. On removal, the oesophagus was filled with fluid and none escaped by the lower end. After a catheter had been passed water ran through but an hour later the cardia was again impermeable. Numerous sections of this oesophagus were examined (1) midway - 10 c.m. from the cricoid (2) above the diaphragm (3) below the diaphragm.

(1) Microscopic examination showed some overgrowth of the epithelial/

epithelial layers and hypertrophy of the muscularis mucosae. The circular and longitudinal layers were also hypertrophied, the latter not so definitely, and some fibrous changes were noted in the muscle fibres. The ganglia appeared to be normal with fairly well stained nuclei and showed no change that could not be accounted for by post-mortem decay.

(2) Above the cardia, similar conditions were found in the muscle layer, the circular coat being again unduly hypertrophied but the epithelium was in great part destroyed and dense masses of leucocytes endothelial cells and fibroblasts had invaded this layer and the subjacent one. The ganglia were surrounded by fibrous bands, showed proliferation of the cells of the capsule and destruction of some and degeneration of other nerve cells.

(3) Below the diaphragm showed less uneven hypertrophy but the circular layer was still by far the most affected and fibrous changes were also more evident and advanced. An occasional nerve cell was to be found staining in a hazy chromatolytic way, but the majority had been destroyed by the encroaching fibrous bands.

(Fig. 5, 6.)

Case III. Oesophagus from woman of 56.

This woman had suffered from dysphagia for seven years and could attribute it to no obvious cause. On admission to hospital she stated that whatever she took was brought up at once and this was confirmed during her residence. All solid or semi-solid foods were at once regurgitated and latterly fluids too were returned.

Under general anaesthesia she had the cardia dilated by Gottstein's instrument and improved considerably after this but died from pneumonia.

This oesophagus differs from nearly all other recorded examples of oesophagectasis by the fact that in no part was hypertrophy of the wall found. The explanation of this is suggested by the microscopic examination.

The upper part of the oesophagus, which measured 8½ inches from the cricoid to the cardia, was of normal calibre. 3 inches below the upper level the dilatation commenced, and assumed pyriform shape, narrowing at the cardiac orifice. In length the dilated area was 5 inches and the width of its widest part was just on 3½ inches. There was practically no variation in the thickness of the wall, 4 m.m. at the upper undilated part, 5 m.m. at the maximum dilatation, 4 m.m. at the cardia. The mucosa was intact, as far as naked eye examination revealed, throughout the specimen, except for some punctate erosions of the cardiac canal.

Microscopic/

Microscopic preparations which were rather unsatisfactory because of the brittleness of the tissue, were made at three levels:-

(1) Through area above dilatation.

(2) Through wall at widest dilatation.

(3) Through cardiac canal.

(1) Microscopic sections showed that the muscular walls had undergone considerable fatty and other degenerative changes. The ganglia were recognisable although a considerable deposit of fibrous tissue was found. Many inflammatory and endothelial cells were distributed over the sections which were shredded and fragmented and looked as if the tissue was beginning to show disintegration.

(2) Showed little alteration from the above changes, no epithelium was seen, nearly all the submucous layer seemed to be desquamated, but nerve cells could be still recognised in the intermuscular zone.

(3) Showed denudation of the epithelium with invasion of leucocytes here and there in deep aggregated patches. The muscular layers were in an advanced stage of degeneration, parts being replaced by fibrous tissue, others showing advanced fatty and hyaline change. The nerve ganglia were absent in many sections and were replaced by fibrous tissue. When the remains of a ganglion were seen, only an odd cell was discernible and inflammatory cells surrounded it. Fig 8.

In/

In this example I have little doubt that we were looking at the terminal phase of an hypertrophied muscular wall. Although the degenerative muscular changes were advanced, the area occupied by the respective muscular coats had not sensibly diminished, from what would be found in a normal specimen and one could come to no other conclusion than that had this oesophagus come to postmortem examination three or four years earlier a very different picture of the thickness of the muscle wall would have been presented.

(Figs. 7.8)

Case IV. - Oesophagus from a woman of 50 years.

This patient had suffered from dysphagia and regurgitation for four years. No complete dilatation of the cardia had been performed but bougies had been passed whenever the symptoms became so acute that swallowing was almost impossible. She died from a broncho pneumonia following influenza.

The oesophagus from the cricoid level to the hiatus measured 22 c.m., the abdominal oesophagus measured 2 c.m. in length.

From the cardia an area of dilatation extended to within 6 centimetres of the upper level fusiform in type. The wall at the top of the oesophagus measured 4 m.m. at the area of maximum dilatation 6 m.m. and in the cardiac canal 5 m.m. On opening the oesophagus milk and particles of bread were found and the lower end of the oesophagus showed patchy pitting of the mucosa. Microscopic sections through the dilated part showed hypertrophy of all the muscular coats but especially the middle, and fibrous and fatty changes were found in some of the muscle bundles. The plexus seemed to be normal.

Sections through the wall below the diaphragm revealed hypertrophy of the muscular coats but not to such degree as was noted above, nor were the muscle fibres so healthy looking, very considerable fibrous change having occurred and in some ten sections the ganglia were entirely replaced by fibrous tissue. When ganglia were seen they were surrounded/

surrounded by fibrous tissue and considerable chronic inflammatory changes were noticed.

(Figs. 9.).

Case V.

In this case and the following one I am indebted to Dr Irwin Moore for the opportunity of making microscopical examinations.

Oesophagus from a man of 60 years.

The specimen was taken from a man aged 60, who had suffered from dysphagia for seven years. He had little difficulty in swallowing fluids, but solids required an interval of from five to ten minutes, and an average mouthful was usually regurgitated.

For twelve months he had been losing flesh, and had developed a cough with abundant and offensive sputum. A bismuth meal remained in the lower end of the oesophagus for 25 minutes.

Death was due to suppuration from a unilateral bronchiectasis.

The oesophagus fig 10 removed in its lower 6½ inches showed a slight/

slight fusiform dilatation measuring 1¾ inches longitudinally and 2½ inches transversely. It commenced at the upper surface of the diaphragm where its calibre was 1 inch. The mucous membrane was normal except at the lower end where there was slight superficial erosion; it was freely movable over the subjacent muscle. There were no ulcers or cicatrices at the lower end of the oesophagus or in the adjacent portion of the stomach. The muscle was hypertrophied except in the last half inch of the specimen. At the upper part the muscle measured 1/10" in thickness, ½" at the area of maximum dilatation and 1/10" at the lower dilated portion. There was no localised hypertrophy corresponding to a sphincter.

Microscopic Examination. (Cardiac Canal).

The epithelium is present but in most places the superficial layers are shed; in one or two places there appears to be increase of cells. In both muscular layers there is hyaline degeneration of the fibres and vacuolation is to be seen in parts; some of the bundles appear to be reduced in size. There is considerable fibrous change around the ganglia and the nerve areas seem contracted with the cells glassy looking and structureless.

Some inflammatory change was found in the submucous tissue and the vessels were thickened and seemed rather numerous.

(Fig. 10,)

Case VI. Oesophagus from a man of 44 years.

This specimen was taken from a man who had difficulty in swallowing since he was 16 years old. He was first seen nine years before his death and then his condition was so advanced that no instrument could be passed into his stomach from above.

Gastrostomy was performed and the cardiac orifice was dilated by the fingers; some temporary relief followed. Ultimately, however, the patient had great difficulty in swallowing, even fluids passing very slowly into the stomach. He became extremely weak and emaciated and died from starvation.

Post-mortem Examination.

fig 11.

The oesophagus was grossly dilated in its entire length except its diaphragmatic portion and upper extremity and formed a large S-shaped bend. At its upper end the normal calibre of the oesophagus began quickly to show signs of dilatation and about its middle reached a diameter of 3 inches. Below this it again gradually diminished in width and was of normal size at the diaphragm.

The dilated portion formed a definite curve with the convexity to the right. Except in the sub-diaphragmatic portion and at the upper extremity the muscular coat of the oesophagus was hypertrophied but was thinned out again in the part forming the convexity of the curve. The

hypertrophy was most distinctly marked below the level of the diaphragmatic opening. No cicatrices or ulcers were present in the lower end of the oesophagus or in the adjacent part of the stomach. The lining of the oesophagus was smooth and appears indeed rather thinned out in places. The sac was at least three times the size of the stomach, which was small and atrophied, and a thick finger easily passed through the cardia.

Microscopic Examination of sections made just below the diaphragm showed general denudation of the epithelium with the submucous layer thickened and fibrosed and densely infiltrated with leucocytes.

The muscular layers, mucosal, circular and longitudinal were hypertrophied and showed also a good deal of fibrosis. Around the ganglion cells some leucocytes, plasma cells and endothelial cells were found and the capsule of the ganglia was thickened and infiltrated. A few nerve cells were to be seen, but they were chromatolytic and ghost like.

(Figs 11, 12)

Case VII. Oesophagus from a man of 70 years.

This patient was admitted to hospital as a doubtful case of gastric carcinoma. There had been a long history of vomiting immediately after meals; occasionally the vomited material was said to be tinged with blood, and acute pain was complained of radiating from the lower end of the sternum over the left side of the chest.

He was only in hospital three days when he died during an anginal attack. At the autopsy there was considerable dilatation of the left side of the heart, degenerative changes in the heart muscle and atheroma of both coronary vessels.

The stomach was small in size and from the cardiac orifice the oesophagus formed a moderately wide dilatation about 9 c.m. in length and 7 in. width. No very marked hypertrophy of the wall was evident, but the thickness was a little greater than normal in the dilated part measuring nearly 6 m.m.

No cardiac canal in the ordinary sense of the term was apparent, the dilated oesophagus meeting the stomach in an abrupt narrow neck.

Microscopic examination just above the neck revealed the customary hypertrophy of the circular layer with less definite overgrowth of the longitudinal. Around the nerve plexus and through the submucous layer numerous inflammatory cells/

cells were found and the ganglionic cells were degenerated and surrounded by fibrous tissue.

A section through the oesophagus nearly at its middle showed very slight inflammatory changes but there was considerable degenerative change in the muscle layers, the circular appearing to suffer most.

Case VIII. Oesophagus from a woman of 30.

In this patient dysphagia had commenced when she was four months pregnant. Soon after the birth of a healthy child obstruction had become complete and gastrostomy was performed. She learned to feed through the tube but later died.

From the cricoid level to the cardiac orifice the oesophagus measured 22 centimetres and presented a fusiform dilatation from the cardia upwards for 13 centimetres and measured 6 c.m. at its widest.

There was slight hypertrophy of the wall of the epicardia and this gradually increased until the level of greatest width was reached, whence it thinned again to normal.

Three levels were examined microscopically.

(1) Upper inch of oesophagus.

No change of any kind was revealed from normal appearances except that the muscular coats showed slight degenerative/

degenerative changes.

(2) Middle of dilatation.

Also practically normal apart from the increase of the muscle fibres and chiefly in the circular coat, and there was also some inflammatory change in the submucous tissue.

(3) Section through the cardiac canal showed a marked degree of fatty and fibrous change in the muscular layers, hypertrophy of the circular layer and the muscularis mucosae. There were inflammatory changes of a not very striking character throughout the specimen except in the intermuscular layer where the nerve cells had suffered extensively. An occasional cell had been spared but generally the plexus showed replacement by inflammatory cells, and evidence of continued inflammatory change in their surroundings. As the photograph shows, complete bundles of muscle fibres are in parts replaced by fibrous tissue.

(Fig. 13)

In each of these eight examples an inflammatory lesion of greater or less chronicity was found in or around the ganglia which control the musculature of the cardiac canal. But in none is any light thrown upon the inception or development of the inflammatory process, although the existence of fibrous tissue with occasional inflammatory cells usually is presumptive evidence of an earlier acute or subacute lesion.

In the following example and the last of this series the sequence of developments is more evident and in my opinion demonstrates clearly the origin and production of this pathological process.

Case IX. Oesophagus from a boy of six years.

An emaciated male child of six years was brought to the Victoria Infirmary, Glasgow, with the complaint that since an attack of broncho pneumonia at eight months, difficulty had been experienced in feeding him. At that time he began to vomit immediately after food, but intervals of from ten days to three weeks occurred in which this did not happen, and once for a period of six weeks he was free. For four weeks before admission he regurgitated every kind of food and drink which he was given and for the last few days of that time would not even attempt to swallow. Shortly after admission to hospital he died mainly from starvation/

starvation although a gastrostomy had been performed in a last effort to give him nourishment.

Post mortem examination.

The only points of interest in the autopsy protocol were that a scar of healed phthisis wasfound at the apex of the right lung, congestion was present at the right base and one or two of the bronchial glands were calcified. No evidence of active tuberculous infection existed and the pyloric and pelvi-rectal areas were of normal size and appearance.

The oesophagus measured 13 centimetres in length from the cricoid to the cardia and showed a fusiform dilatation 5 centimetres in width at its maximum. At the cardia the measurement was just over 1 centimetre and there was manifest hypertrophy of the wall of the cardiac canal, and was greater than anywhere else measuring 6 millimetres, in the dilated part between 4 and 5 m.m. and at the top of the oesophagus just over 1 m.m.

Several minute erosions were obvious to naked eye examination throughout the affected area.

Microscopical Examination of sections through the cardiac canal showed the epithelial layer to be pitted and in other places denuded. Intense inflammatory zones formed the bases of these shallow ulcers and leucocyte and lymphocytic invasion around the engorged vessels was extreme.

The/

The muscularis mucosae was increased in thickness while hyaline changes were observed in many of the fibres. There was striking hypertrophy of the circular muscle coat and here and there, inflammatory cells occupied the interstices; degenerative changes fibrous fatty and hyaline were seen. In the longitudinal layer such evident change was not in progress though these films also showed degenerative change with a scattering of leucocytes and endothelial cells. In the intermuscular layer the exudation of leucocytes, plasma cells and endothelial cells was abundant. The ganglia as a whole were affected and presented in places, a glassy hyaline appearance with only one cell or perhaps two showing a nucleus while the capsule was lined and surrounded by inflammatory cells. Again, an area of what appeared to be colloid presented where nerve cells would ordinarily be expected. The capillaries in this area were congested and their endothelial cells were swollen. Despite the most careful search no tubercle bacilli were found but pneumococci were seen especially in the intermuscular zone.

(Figures and Drawings.) 14, 15, 21.

In this example, the inflammatory zone stretched unbroken from the mucous membrane to the tunica adventitia, and was acute in character. If this child had lived and had been successfully treated, the regression we should ultimately have expected would have been manifest in subsidence

of/

of the inflammation, the laying down of fibrous tissue probably where inflammatory changes were furthest advanced and the removal, partial or to a great extent, of the damaged structure by phagocytosis.

In such circumstances, little evidence of the damage might be found in the relatively lowly developed submucous or muscular layers, but in the more highly organised nerve cells permanent evidence of destruction would be found. The muscle bundles too though hypertrophied at the height of the process would probably undergo similar alterations in size when restitution of tissue commenced, so that an originally hypertrophied muscle might appear of normal or even diminished thickness.

This I take to be the ordinary development of the chronic or subacute lesion in the eight cases already described.

It is of course possible that similar effects might be produced by the specific selection of ganglia by circulating toxins: not only is this paralleled in other conditions, but it would account for those cases of oesophagectasia which would appear to have followed an acute febrile disease. But even in the specific fevers and after influenza, ulceration of the oesophageal mucous membrane, or short of such severity an acute inflammation, is sometimes found and in no part of it is ulceration more likely to occur than at its lower/

lower end where stasis of short duration is always found. An outward spread would eventually involve the ganglia with cessation of local peristalsis.

As no Meissner's plexus is found in the gullet, it follows I think that its functions must be subserved by Auerbach's plexus, which not only controls the peristaltic wave but receives stimuli from the nerve endings in the mucosa. The peristaltic wave can be started at any point in the oesophagus and a lesion affecting the nerve endings alone or the fibres at any point in their course between the epithelium and the ganglia, might interfere with peristalsis long before the ganglia themselves showed any demonstrable histological lesion. It may be as well here to refer to a peculiar case which came to routine post-mortem examination, and is not included in this series because it seems to me to belong rather to the category of rumination or merycism.

The oesophagus presented a saccular dilatation extending about 4 or 5 inches from the cardia and about three inches wide. There was no alteration in the thickness of the wall or the muscular coat anywhere, nor was there any obstruction or constriction anywhere. The oesophagus tapered at the cardia and no canal in the ordinary sense of the term was visible and the stomach showed a similar indipping about half way along the

lesser/

lesser curvature. The stomach itself was of fair size but certainly not enlarged. The Pylorus appeared to be normal.

The sections made from the wall of the oesophagus (in its dilated portion) were not satisfactory but they showed that the muscular layers were not increased; there was practically no epithelium, the submucous coat showed a few thick blood vessels and fibrous changes throughout the specimen were advanced. The ganglia were altered but not more than the rest of the tissue and the change was probably atheromatous in origin.

He was a fisherman in the West coast of Scotland and was in hospital for cardiac disease of old standing. One of his friends said that he had always been "funny about his food" and "could bring it up like a cow", but he did not think that he ever vomited. This of course may have been a dilatation of the oesophagus similar to the cases just described from primary spasm or achalasia. Even in life it would probably be a difficult enough case to diagnose and as I have said I believe it to have been an example of rumination, though I can find no similar case mentioned in the relevant literature. He had also a horseshoe kidney and considerable dilatation of the aorta.

Dilatation of the oesophagus in this affection follows three main types -

(1) Fusiform

(2) Flask or pear-shaped

(3) S shaped.

(1) The spindle shaped or fusiform type is the most common form of dilatation and the maximum width of the oesophagus is usually just below the middle. Obviously in this type the cardia is the most dependent part and unusually with this type there is a cardiac canal of definite length.

(2) In this form the saccular dilatation seems often to bulge over the diaphragm and there is widest; sometimes it forms a sulcus round the opening through the diaphragm which is somewhat raised. In this type also the cardia is the lowest portion. Often a definite cardiac canal is not so obvious, the junction from oesophagus to stomach being the lowest part of the dilated viscus which forms a neck.

(3) The S-shaped variety is extremely uncommon and one has the impression in this example, of the cricoid and cardia acting as fixed points between which the oesophagus lengthens and curves. The curve usually is towards the right and the dilated oesophagus rests on the diaphragm posterior to and to the right of the cardia;

from/

from here it passes upwards, forwards and to the left and enters the abdomen through the diaphragm at a higher level; consequently a reservoir is left behind the level of the cardia.

In this form of oesophagectasia lengthening always takes place. It is a moot point whether in the other types increase in length is usually found. My own experience is that it is not usually found, as comparison of the measurements of twenty normal oesophagi already tabulated with the fusiform examples in this series, will show.

Sometimes there is quite a distinct backward bend at the lower end which leads to the oesophagus meeting the stomach at rather an acute angle. In these there is probably lengthening too.

The form of cardiospasm which I have been describing and which I believe usually follows a primary oesophagitis, might be called direct cardiospasm in contradistinction to referred or reflex cardiospasm which may accompany spasmodic contraction of the pharyngo-oesophageal sphincter or gastric or duodenal ulceration.

In these conditions, there is often a delay in the passage of a barium meal or drink through the cardia, but I have not yet come across an example in which the

apparent/

apparent hypertonus was severe enough to produce oesophagectasia.

Age and Sex Incidence.

The sexes appear to be about equally affected as far as the limited statistics show. I have seen 20 cases, of whom 11 were men and 9 women; and it would seem to be a disease of middle age though some early cases have been reported. The child in the present series is the youngest that has come to post mortem examination as far as I can find. Langmead reports a case in a child 18 months old, where the symptoms commenced shortly after birth; Chevalier Jackson, a case in a child two days old; Morgan collected 5 cases occurring in children under 2 years old and Parkes Webber, Gopport and Roziere each report one similar case, in slightly older lesion. These appeared to be definitely examples of cardiospasm, as judged by X-ray and endoscopic examination and not secondary dilatation following congenital hypertrophy of the pylorus. A very recent case reported by Van Gilse commenced shortly after birth and appeared to follow birth trauma, a copy of the suggested lesion is given (Fig. 16.)

The symptoms are essentially dysphagia and regurgitation with debility and loss of weight.

Dysphagia may exist in almost any degree, from a slight difficulty,

difficulty in getting solid food into the stomach, to an inability to swallow even fluids without regurgitation; and the obstruction may be momentary or transitory; on the other hand it may be of long duration or complete. Some patients are able to force the food onwards by swallowing movements or with the assistance of large quantities of fluid. Others by a series of deep breaths appear to clear the obstruction and some find relief by stretching or jumping up and down, or by assuming unusual positions such as inclining backwards with the legs fully extended.

Often the onset of dysphagia is insidious and may in its earlier stages excite very little attention if any at all, only when regurgitation is superadded does the patient realise that serious trouble has begun. The latter symptom is inevitable as soon as dilatation of any degree is established, unless a pari passu and compensatory hypertrophy of the muscle wall has been established.

Regurgitation comes on readily and often with no effort immediately after rising from table or while fasting, and may be especially troublesome after going to bed. Blood may be found in the regurgitated material but naturally no gastric juice, and it is often because of this fact that the patient's attention is drawn to the difference between this "vomiting" and ordinary sickness in which the gastric contents are evacuated.

The amount of regurgitated food is, in advanced cases,

often only an ounce or two less than that swallowed, so that soon considerable wasting develops.

Pain is an inconstant symptom and when present may be radiated over the epigastric region or be centralised at the lower end of the sternum. It may "go through to the back" or it may be radiated over both arms and shoulders. More often a sensation of fullness or heaviness or dragging in the epigastrium is complained of and then at the end of a meal and relieved frequently by regurgitation. Salivation is often excessive, especially at night time. In time there is weakness, cachesia and rapid wasting.

Differential Diagnosis.

Although in arriving at a diagnosis of cardiospasm, consideration must naturally be given to extra oesophageal pressure from mediastinal tumours and similar conditions, the effects of corrosives and impacted foreign bodies, the essential differentiation is from cancer of the epicardiac canal or cancer high up in the stomach wall. The diagnosis is obviously of the first importance, is sometimes most difficult and occasionally impossible. There is never invasion of the epicardiac canal by tumour without some degree of spasm being superadded, and although earlier in this paper I drew attention to the fact that in cardiospasm, very often the muscle higher up appeared to be contracting to the limit of its powers, and in carcinoma of the lower

end/

end of the oesophagus this is not seen; this distinction is obviously dependent upon the amount of spasmodic contraction which may accompany the malignant infiltration.

X-ray examination may be otherwise helpful. In cardiospasm, below the dilated area there is often a threadlike constriction fading suddenly into the anterior wall of the stomach, the edges of the curved sides as they approach to the constriction are usually clean cut (Figs. 22, 23.) and meet evenly; one side is not usually higher or larger than the other. There is no suggestion of infiltration of the lips, nor is there darkening or shadow round the lines of the constriction, as one usually finds in cancer (Figs. 24, 25.) whether epitheliomatous or in the submucous coat. The dilatation in cardiospasm is seldom so limited as in carcinoma.

Endoscopic appearances usually settle the diagnosis. The presenting of an ulcerated, papillomatous or mamillated surface on examination puts the diagnosis beyond doubt; but if the malignant tissue is confined to the submucous coat and the epithelium is unbroken, no help at all from oesophagoscopic examination may be given. Such cancers are often those in which the greatest degree of spasm is found, and they may be found just above the diaphragm, in the diaphragmatic part or below, when they often are

extensions/

extensions from a neoplasm on the stomach wall. Sometimes the appearance of a crescentic or reddened patch on the epithelium may reveal the malignant process; at other times tacking down of the epithelium or involvement of the muscularis mucosa may leave a patch which on endoscopic examination is relatively at rest during the respiratory movements.

If any doubt should be entertained after X-ray and oesophagoscopic examinations, it is probably wise to perform gastrostomy and leave the diagnosis to digital examination. The following case illustrates the necessity for this.

A man of 45 who was admitted to hospital with dysphagia and regurgitation was confidently diagnosed as cardiospasm on X-ray and endoscopic examination (Fig. 26) Bougies were passed and two months later he was again back in hospital, if anything much worse (Fig. 27). On this occasion, gastrostomy was performed and high up in the stomach wall and close to the cardia an adeno carcinoma was found. The microphotograph shows the invasion of the area round the ganglia by inflammatory and tumour cells. (Fig. 17). The invasion of the ganglionic surroundings no doubt caused the intense spasmodic contraction, and diagnosis of such a case could not be made from either X-ray or endoscopic examination and gastric analysis could not be performed.

Paralysis/

Paralysis of the oesophagus, a very rare condition, may present symptoms not unlike those of obstruction at the cardia. Diagnosis here is said to be established by the ease with which the walls of the oesophagus fall apart on examination and if a capsule is introduced well into the oesophagus, it will be driven on by peristalsis if the muscle is acting, but will remain stationary if there is paralysis.

Treatment.

No medical treatment, though many drugs have been employed, has been found to be of any lasting benefit. Some method of dilatation must be used and this may be either instrumental or digital.

Among the former many varieties are to be found but they are essentially of two kinds, either a modification of the original dilatable rubber bag introduced by Russell of Southport in 1885, or a bougie weighted with mercury.

Sippy's, and Gottstein's are the two probably most commonly used in this country; Mosher's and Jackson's modifications in America. Hurst's mercury tube has now had an extensive trial and has appeared to give good results, although there would appear to be obvious dangers in the indiscriminate use of this instrument - it contains 1 lbs. 6 oz. of mercury - by the inexperienced or by those who are unaware of the frequency with which cardiospasm may be confounded with carcinoma/

carcinoma of the cardia.

For digital examination gastrostomy is a necessary preliminary and care is required in stretching the cardia.

Whichever method is employed, and each has its enthusiastic adherents, the essential thing is to obtain a free and wide passage by the greatest practicable dilatation of the cardia.

Results.

The immediate effects, the pain and discomfort due to manipulation having passed away, are usually excellent. It is extremely difficult, however, to be confident of the ultimate results. For a period of certainly two years, and sometimes longer, one dilatation seems often to be sufficient. But there is an unquestionable tendency for recurrence to take place, although this may be again relieved by the instrumental aid.

I have attempted to investigate the rather interesting question, does the dilatation of the oesophagus remain after successful treatment? And to this I can only so far give the guarded answer, that for three months afterwards, it appears to remain dilated. I have only had the opportunity of "following up" three cases and in each so rapidly did the Barium traverse the oesophagus, and from the transient glance afforded, so wide did the shadow appear, that any other conclusion was inadmissible.

In/

In successful cases, doubtless as time passes, a balance is struck between the amount of dilatation and the extent of the hypertrophied wall but at the best this equipoise must be precarious, as fatty and fibrous degeneration will tend to replace the muscle fibre when the necessity to overcome the contracted cardia no longer exists. If, in addition, the mucous membrane is constantly suffering from the effects of carelessly masticated or bolted food, it is hardly to be wondered at, if dysphagia returns from inability of the exhausted muscle of the oesophagus to force on its contents, although the cardia may be even of wider limits than normal. I have not seen such a case, but one has been reported by St.Clair Thomson.(22)

In such a case, where there is nothing to dilate, all that could be done would be to make a trans-peritoneal anastomosis between the dilated oesophagus and the fundus of the stomach. On one occasion at least, this was successfully tried by Grey Turner.(22)

***
*

REFERENCES.

The following, chiefly English and American references, deal with the modern development of this subject. The earlier French and German references are to be found in Brown Kelly's and Walton's papers, Nos. 1 and 29 in the list:-

---

(1) Brown Kelly, A:- Brit.Med.Jour. 1912. Vol.ii.Oct.19th.

(2) do. :- Proc.Royal Soc.Med., Lond. 1919, Vol.xii, p.48.

(3) do. :- do. xiii. p. 206.

(4) do. :- Journ. Laryn. and Otol., Edin. xlii, 4, 1927.

(5) Cameron, J.A.Munro:- Arch. Dis. Child., Lond, 1927, ii, 12.

(6) do. :- Jour. Laryng. and Otol., Edin., xliii, 3, p.218.

(7) Hill, Wm. :- Jour. Laryng. Otol. Edin., 1912, xxviii, 2.

(8) Hofer and Spiegel:- Monatschr. f. Ohrenheilk. v. Laryng. lviii, H.8.

(9) Hurst, A.F.:- Brit.Med.Jour., 1925, i. p. 145.

(10) do. :- Lancet, Lond., 1927, i. 618.

(11) Irwin Moore:- Journ.Laryng. and Otol., Edin., xlii, 9, p.577.

(12) do. :- Proc.Roy. Soc.,Med., xii. p.67.

(13) Jackson, Chevalier:- Peroral Endoscopy Endoscopy and Laryngeal Surgery - 1915.

(14) Jackson, Chevalier:- Bronchoscopy and Oesophagoscopy, Saunders, Philadelphia.

(15) Jacques and Rousseaux:- Acta. otolaryngol. xii. H.1/2.

(16) Lambert:- Surg. Gyn. and Obst., 1914, xviii, p.1.

(17) Mosher, H.P.:- Pensylv.Med.Jour., 1912.

(18) do. :- Amer.Jour. Oto-Laryngol., 1928.

(19) Pritchard, E. and Hillier, W.T.:- Proc.Roy. Soc.Med. (Child), xiii, 33.

(20) Parkes Welber, E.:- Proc.Roy.Soc.Med., (Child), xiii, p.36.

(21) Rake, G.W.:- Guy's Hosp. Reps., Lond., lxxvi, Z.

(22) Royal Society of Medicine:- Discussion on Cardiospasm, Oesophagectasia. Sections of Laryngol. and Dis. of Child., 1912, 1919, 1926, 1927, 1928.

(23) Stokes, A. and Rake, G.W:- Guy's Hosp. Repts., Lon., lxxvii, p.141.

(24) Russel, J.C.:- Brit.Med.Jour., 1898, i. p.1450.

(25) Thomson, St.Clair:- Proc.Roy.Soc.Med. Discussion, Vol.xii, p.38.

(26) van Gilse, P.:- Zeit. Hals. Nasen. Chrenheilk, Berlin, 1928. Bd.22, 4. 1.

(27) Walton:- Brit.Jour.Surgery, Bristol, 1925, xii, 48.

***
*

THE PELVI-RECTAL SPHINCTER,

Sphincter of O'Beirne.

At the transition from the colon to the rectum, there is an area in which stasis tends normally to occur, but at which no local thickening of the muscle wall is sufficiently obvious to term it an anatomical sphincter. O'Beirne in 1833 drew attention to the sharp angle which is formed by the junction of the relatively mobile pelvic colon and the fixed rectum, and suggested that prior to the act of defaecation the bowel contents tended to accumulate at this level and not as was previously thought in the rectum. More than fifty years later Hirschsprung described in a child, an extensive dilatation of the colon which appeared to begin at this junction yet did not appear to be caused by any obstruction or natural narrowing. Many examples of megacolon have been reported since 1886 and the disease appears to be more common than was at first believed.

In general terms it may be said that while many cases, probably the majority, date from birth, others appear to develop in infancy or childhood. Some proceed early to a fatal issue; in others a modus vivendi is attained/

attained and apparently active and healthy lives may be led in which the only symptom of pathological disorder is severe and obstinate constipation. Hurst indeed describes a patient who rowed for his University.

Now between idiopathic dilatation of the oesophagus and dilatation of the colon there are such obvious resemblances, that a similar pathological lesion might be expected in both. It is true that there is a decided difference in age incidence, the former being essentially a disorder of the middle years, the latter of infancy and frequently congenital, but apart from this there is no essential difference and I propose to show in the following pages that the pathogenesis and aetiology are identical.

Briefly my thesis is as follows:-

(1) The ultimate lesion in Hirschsprung's disease is a degeneration of the cells in the ganglia controlling the pelvi rectal sphincter,

(2) That in congenital cases the primary factor is bacterial fermentation of meconium in the relatively long and lax pelvic colon; an inflammation of the mucosa is produced which spreads outwards to involve the ganglia.

(3) The involvement of the ganglia causes interruption or undue delay of peristalsis at the pelvi rectal sphincter so/

so that material accumulates and dilatation of the colon follows.

(4) In an attempt to expel the unaccustomed burden, the muscular wall of the bowel hypertrophies, the hypertrophy affecting specially the chief contracting layer, the circular muscle coat.

(5) Once the optimum relationship is attained between dilatation of the bowel and hypertrophy of its wall, a natural cure of the obstruction at the sphincter is attempted and with the subsidence of the acute inflammatory process, restitution of the muscular and mucous layers may begin while the more highly developed nerve plexus cannot take place. A late examination therefore might only show changes in the ganglia while the other layers appeared to be nearly normal.

(6) In those cases which develop months or years after birth the initial factor is probably a lesion of the anus or rectum through which inflammation spreads in the same way. It must of course be understood that even in those, the lesion might still be congenital and symptomless clinically until the compensation between the dilatation of the bowel and the hypertrophy of its wall, broke down.

Anatomy.

The pelvic colon varies in length and tortuosity in any series of specimens; the shorter it is the fewer the bends in its course. Beginning at the medial border of the left psoas major it usually crosses the pelvis from left to right, bends backwards and slightly upwards, and then sharply downwards to become the rectum at the level of the third sacral vertebra. It may proceed almost right over to the right pelvis wall before bending and in the child the greater part of its course is abdominal and not pelvic. The extensive fan shaped mesocolon, short at each end and longer in the middle allows considerable freedom of movement and in the newborn child is relatively much longer than in the adult. The arrangement of layers, glandular, submucous, muscular and adventitia, is similar to that found elsewhere in the intestine, but in the mucous membrane and the longitudinal muscle there are differences between this area and the rest of the colon. In the former there is a tendency for the membrane to be gathered into longitudinal pleats in the last five or six centimetres of its course, very similar to what is found at the lower end of the oesophagus. And the three taenia in which the longitudinal muscle of the colon is generally arranged, merge/

merge together as the pelvic colon is traversed, so that midway only a narrow area of the bowel on each side is uncovered by longitudinal muscle and at the sphincter even these disappear, so that the investment is complete.

No other anatomical differences exist between the pelvi rectal sphincter and the rest of the colon and Auerbach's Plexus is found between the circular and longitudinal coats, and the smaller Meissner's plexus in the submucosa.

Physiology.

The radiological and sigmoidoscopic observations of Hurst and others have confirmed the opinions of O'Beirne, that normally the faeces tend to accumulate behind this sphincter prior to defaecation and that the pelvi rectal junction exercises a sphincteric action weaker no doubt than the anal and pyloric sphincters but comparable to the cardiac sphincter.

Hirschsprung's Disease.

Case I.

A boy aged 8 years was admitted to the Victoria Infirmary, Glasgow, collapsed, and obviously in extremis. Palpation revealed a distended abdomen and death took place while preparation for a laparotomy was being made.

The/

The history obtained from his mother was to the effect that from birth he had suffered from extreme constipation which was periodically relieved by massive stools. At times pain in the abdomen was so severe that the boy had to receive medical attention, but generally pain was not a very common complaint. Until his fatal attack, he had only once been away from school for a week during his two years' attendance.

Occasionally after a long period of constipation he vomited considerable quantities of bile.

The post mortem examination revealed an extreme degree of dilatation of the descending and rather less of the transverse colon, the dilatation beginning sharply at the pelvi rectal junction. The wall of the descending colon was thickened, the hypertrophy affecting principally the circular layer: no abnormal narrowing at the sphincter of O'Beirne, and no adhesion of glands or obstruction by any tumour formation could be seen. The wall of the dilated portion was slimy and the mucous membrane excoriated and sloughing, and there was a general tendency to shed large portions of epithelium.

Microscopic examination of the descending colon confirmed the overgrowth of muscle fibres, circular, longitudinal/

longitudinal and muscularis mucosae, but particularly of the circular layer. Some inflammatory cells were scattered through all the layers, but apart from the hypertrophy no definite pathological change was seen, and the nerve cells stained well and showed no change that could not be accounted for by post mortem decay. Through the pelvi rectal sphincter the conditions were different, and the chief alteration was in the cells of the nerve plexus.

Inflammatory cells were found in the submucosa and through the muscle layers, and around the ganglia; in the ganglia fairly complete degeneration was found, only an occasional nerve cell showing, and staining badly.

Case II.

A boy aged 6 years was admitted to the Victoria Infirmary as an "acute abdomen". Shortly after admission a laparotomy was performed and the peritoneum was found to be blood stained. A volvulus of the dilated colon had taken place and areas of necrosis in the descending part were seen. An attempt at radical removal was precluded by the condition of the patient and a drainage tube was inserted. The post mortem examination revealed a moderately extensive dilatation of the whole colon with some hypertrophy of the wall. The whole bowel was congested and when opened was blood stained with patches of necrosis throughout the pelvic and iliac colon and almost complete sloughing of/

of the mucosa.

The microscopic examination was practically useless as the tissue shredded as it was cut and the most extreme degree of inflammation was found wherever tissue was recognisable.

Case III.

A boy, 7 years old, was admitted as a case of Hirschsprung's disease for medical or surgical treatment. From birth he had suffered from severe constipation and at times the degree of distension of the abdomen was severe. It was comparatively common for him to empty his bowel once in two or three weeks, and during such a prolonged period of constipation he was dull and refused food; he sometimes vomited and once had apparently vomited faecal matter.

On admission peristaltic movements over the distended colon were obvious and an extreme degree of distension was present. Medical measures for relief were tried and with lavage through a long tube he began to show very decided improvement. Later he developed broncho-pneumonia and died.

Post Mortem Examination.

The colon was distended from the caecum to the pelvi-rectal sphincter; its width being five times that of the sphincter. The wall above the sphincter was about four times the thickness of the wall/

wall at the sphincter, and the enlargement was chiefly confined to the circular muscular coat.

Microscopic Examination.

(a) Through the distended colon:-

The mucosa, the muscularis mucosae and the submucous layers all showed some fibrous thickening; the circular muscle fibres were greatly hypertrophied; the longitudinal to a lesser degree. The cells of Auerbach's plexus appeared healthy but some fibrous change was noted in their surroundings.

(b) Through the pelvi-rectal sphincter.

No changes of importance were noted in the epithelial layer and glands, the muscularis mucosae or the submucous layer although a few inflammatory cells were found throughout. The circular muscle coat was in sharp contrast to that in the dilated area above and of nearly normal thickness, as was also the longitudinal layer and showed little alteration apart from some fibrous and fatty degeneration. In the inter-muscular plexus of nerve cells the changes were of a striking character. As the accompanying microphotographs show, the ganglia were replaced by inflammatory cells. In the ganglion which is shown under the high power only one nerve cell shrunken and degenerated was to be seen. (Figs. 18, 19,)

wall at the sphincter, and the enlargement was chiefly confined to the circular muscular coat.

Microscopic Examination.

(a) Through the distended colon:-

The mucosa, the muscularis mucosae and the submucous layers all showed some fibrous thickening; the circular muscle fibres were greatly hypertrophied; the longitudinal to a lesser degree. The cells of Auerbach's plexus appeared healthy but some fibrous change was noted in their surroundings.

(b) Through the pelvi-rectal sphincter.

No changes of importance were noted in the epithelial layer and glands, the muscularis mucosae or the submucous layer although a few inflammatory cells were found throughout. The circular muscle coat was in sharp contrast to that in the dilated area above and of nearly normal thickness, as was also the longitudinal layer and showed little alteration apart from some fibrous and fatty degeneration. In the inter-muscular plexus of nerve cells the changes were of a striking character. As the accompanying microphotographs show, the ganglia were replaced by inflammatory cells. In the ganglion which is shown under the high power only one nerve cell shrunken and degenerated was to be seen. (Figs. 18, 19,)

Comment.

Hirschsprung's disease is therefore exactly comparable in its pathological basis to cardiospasm. Although the lesions that I have described are from comparatively old-standing cases, they are so similar to the chronic cases of cardiospasm which I have already described, that I have little doubt that if a sufficiently early example of mego-colon is obtained the results will be the same as in the acute case of cardiospasm (Case IX) and the complete line of invasion by inflammatory cells will be found stretching from the epithelium to Auerbach's Plexus.

Medical treatment should always be tried at first and the best results obtained so far have followed colonic lavage; purgatives generally are looked on with suspicion; of these some preparation of aloes is likely to be as effective as any and the ordinary methods of combating constipation should be instituted.

Of surgical measures, resection of the affected gut with entero-anastomosis is probably the commonest procedure and the statistics of Schneiderholm showed 46% of cures.

Adson has recently informed me that he has had two recoveries after performing rami-section.

In/

In conclusion, it seems to me that some form of dilatation of the pelvi-rectal sphincter should be given a reasonable trial. The sigmoidoscope and modern X-ray illumination have made blind bougieing of this area no longer necessary, and so successful has dilatation proved in many cases of cardiospasm, that there is no obvious reason why it should not be equally successful in Hirschsprung's disease. The probability indeed is that many of the excellent results that have appeared to follow lavage have been in part, at least, due to the stretching of the sphincter inevitably involved in such manipulation.

References.

Cameron, J.A.Munro:- Arch. Dis. Child. Lond., Aug. 1928.

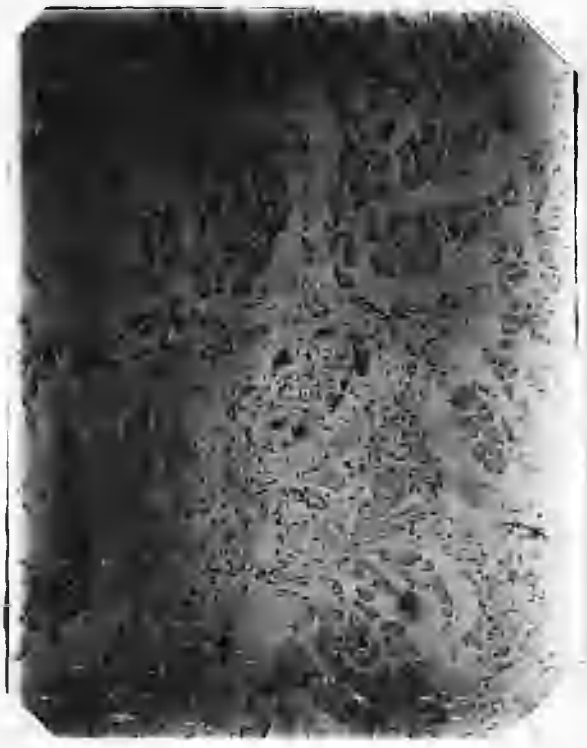
Finney:- Surgery, Gyn. and Obst., June, 1908.

Hurst:- Archiv. de Malad. Nutrition, Jan., 1925.

Schneiderhöhn:- Zeit. f. Kinderh., 1915, xii.

***
*

Fig 1.



Transverse section of œsophagus 2cm. below cricoid

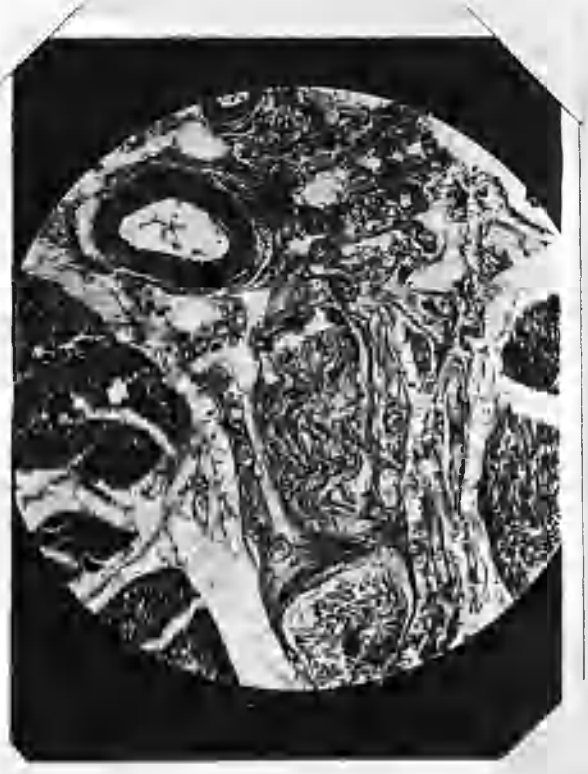
Fig 3.

Fig. 2

Nerve fibres running over circular muscle layer from ganglia

Fig. 4.

Nerve fibres in sub-mucous coat



Through tardiae canal: Fibrosis of ganglia: Thickening of vessel.

Fig. 5.

Shows thickening of cardiac muscle

Fig. 6.

Section through Cardiac Canal

Fig. 7.

Shows no hypertrophy throughout oesophagus.

Fig. 8

Ganglion with degeneration of cells.

Fig. 9.

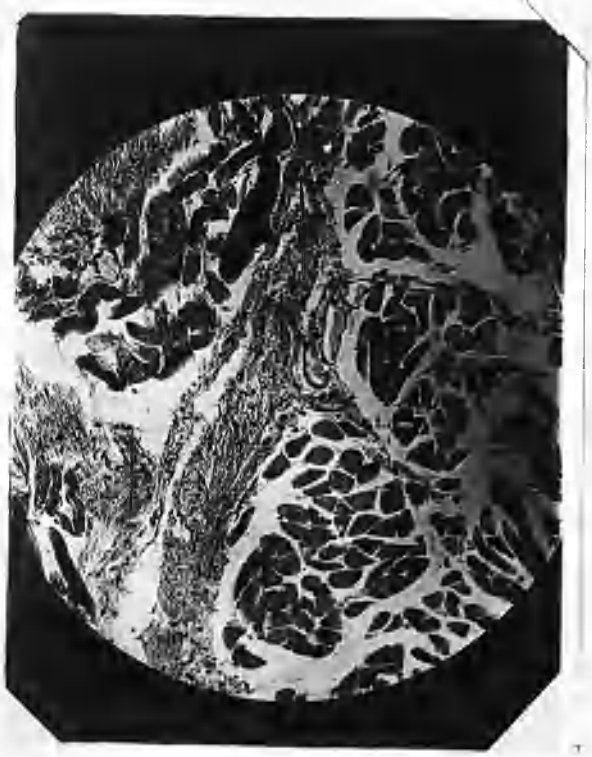


Fig. 10.

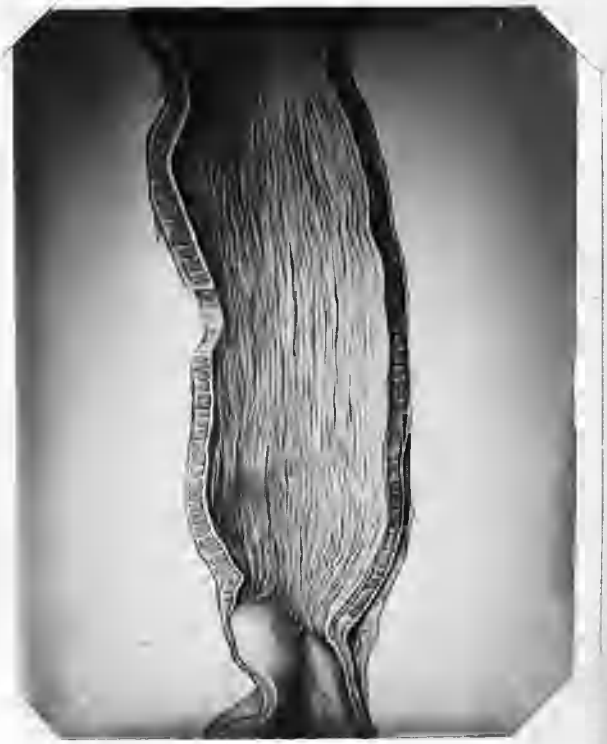


Fig. 11

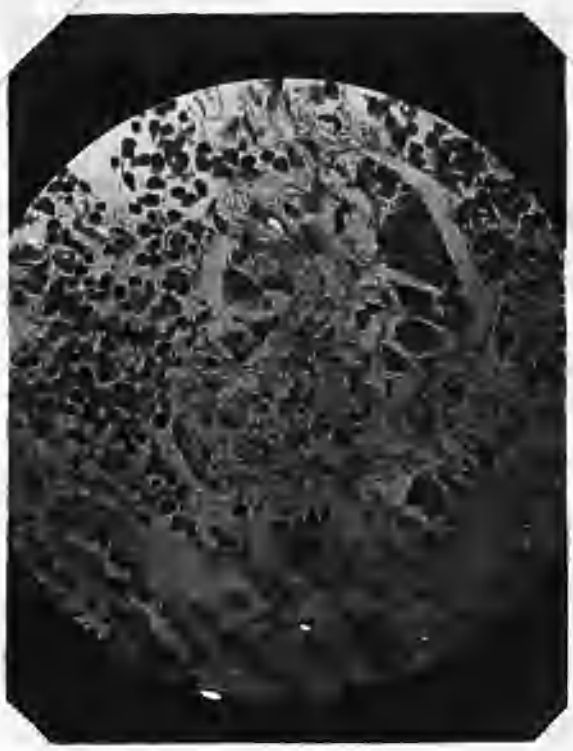
Fig. 12

Showing increase of muscularis mucosæ, and fine separation of muscular

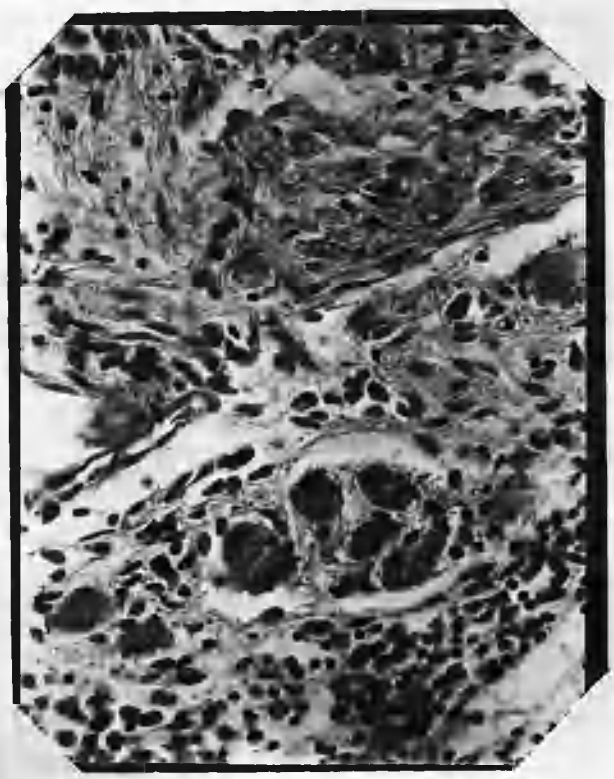
Fig. 13.

Shows advanced fibrous degeneration of circular muscle coat.
Fig. 15.

Fig. 14.



Section through Cardia.
Fig. 16.



Section at diaphragmatic level.

Fig. 17.

Ganglia in stomach wall.
Surrounded and invaded by.
inflammatory and tumour cells.

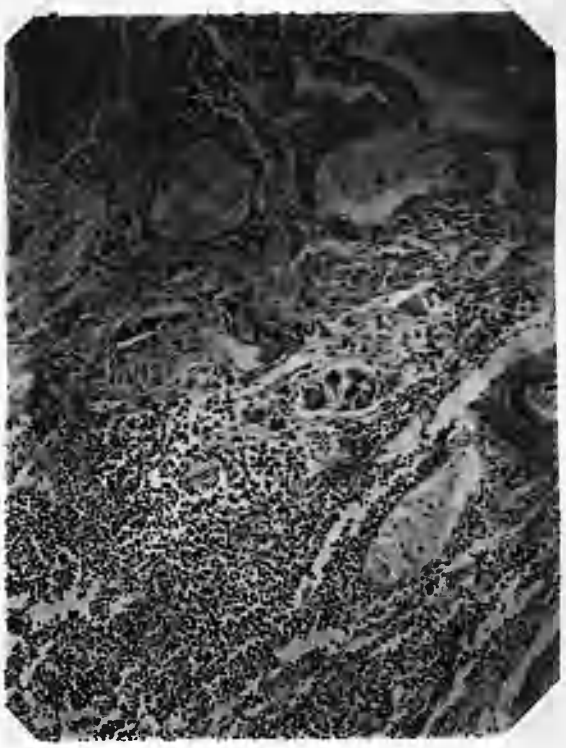
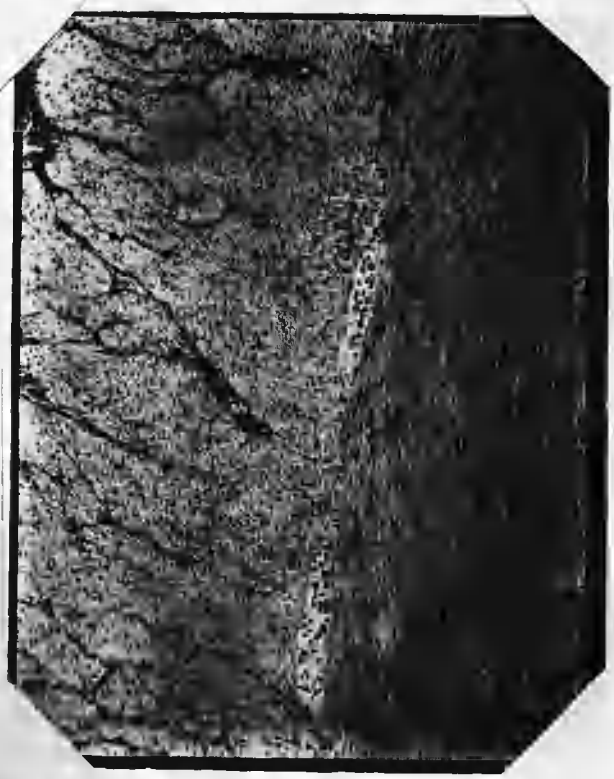
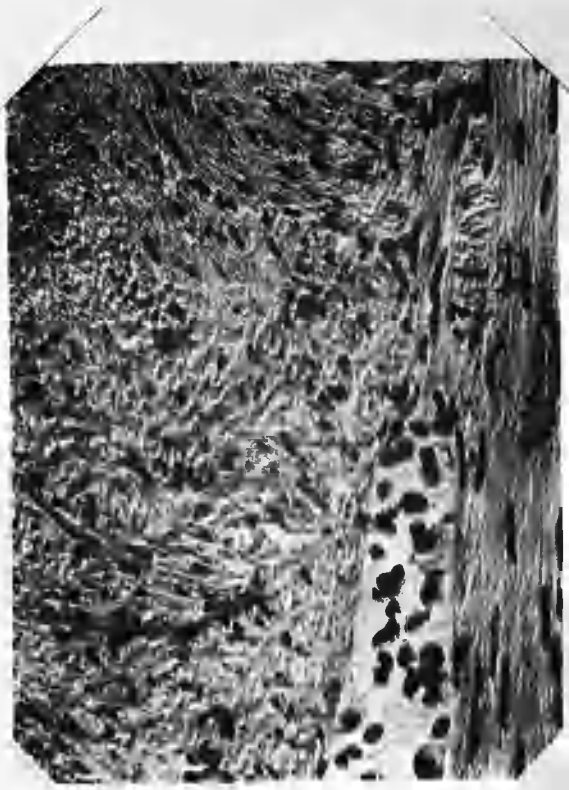


Fig. 18.



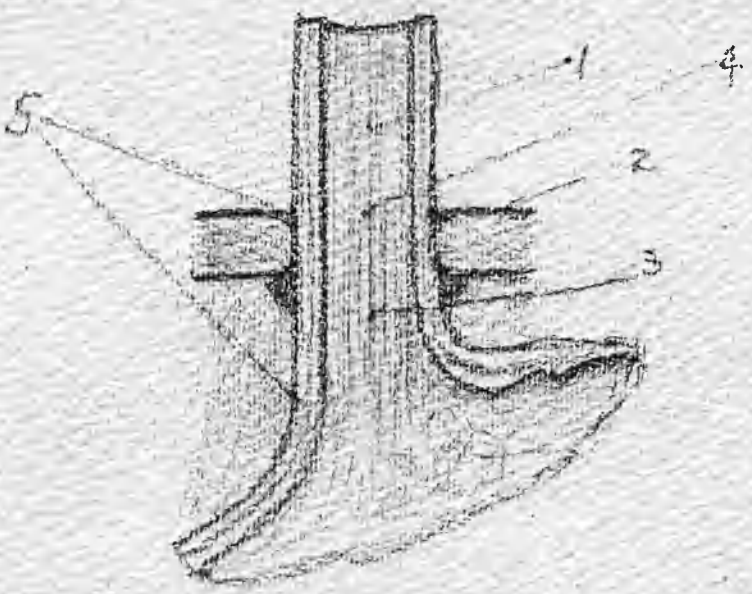
Longitudinal Muscle. Ganglionic layer. Circular Muscle.

Fig. 19.



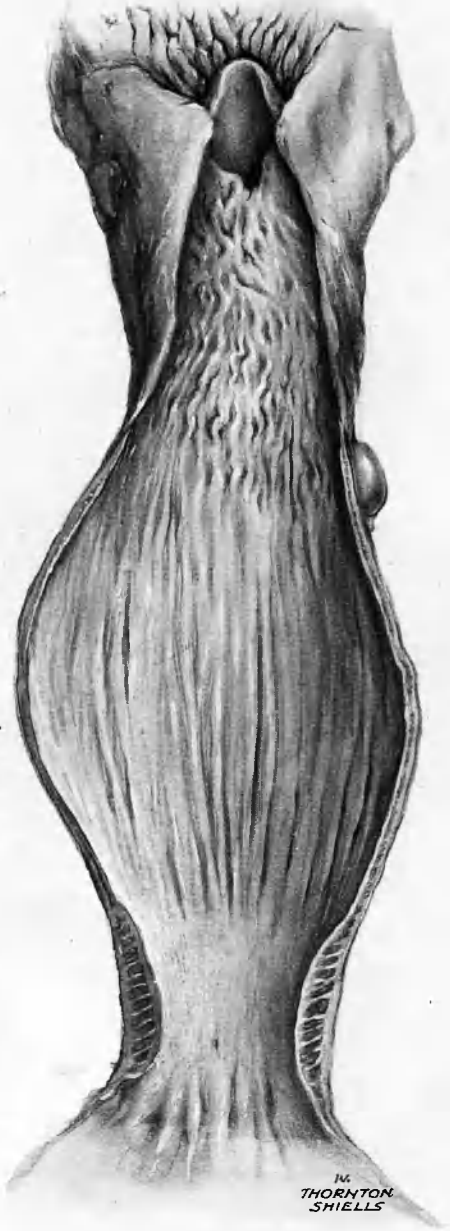
High Power Illustration

Fig. 20.



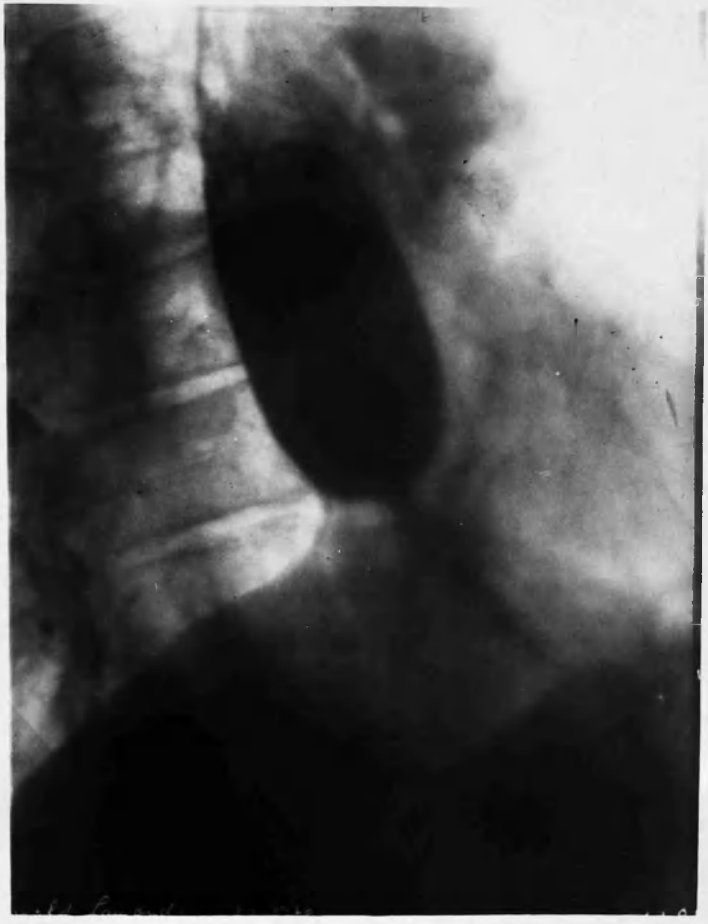
1. Thoracic Oesophagus.
2. Diaphragm
3. Abdominal Oesophagus.
4. Hiatal Oesophagus.
5. Epicardiac Canal.

Fig. 21.



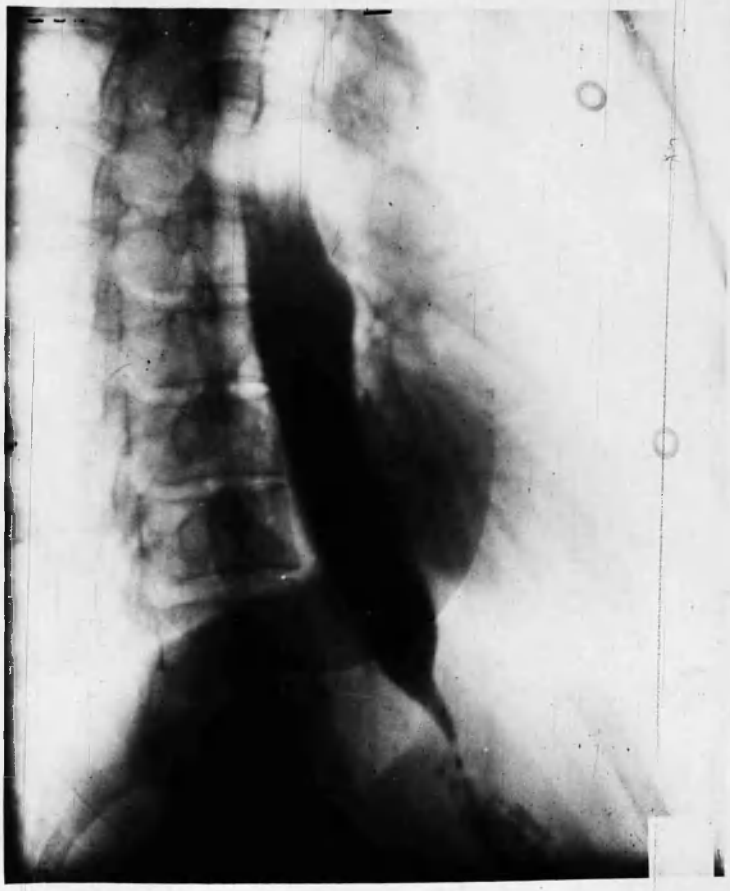
Oesophagectasia. Child — 6 years old.

Fig. 22



X-Ray photograph - Cardiospasm -

Fig. 23.

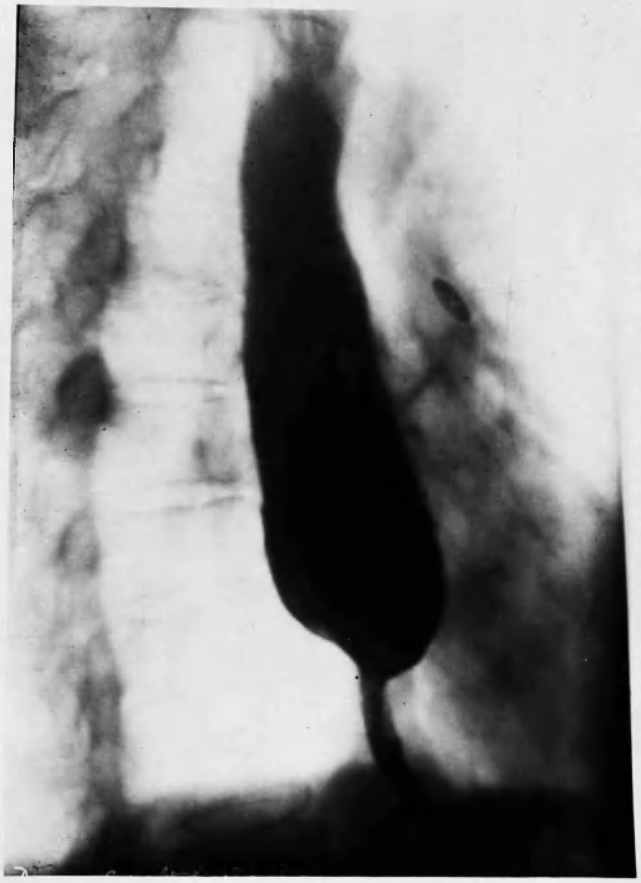


Cardiospasm — X-ray photograph.

Fig. 24.

Carcinoma of lower end of Oesophagus.

Fig. 25



Carcinoma of lower end of Oesophagus.

Fig. 26.

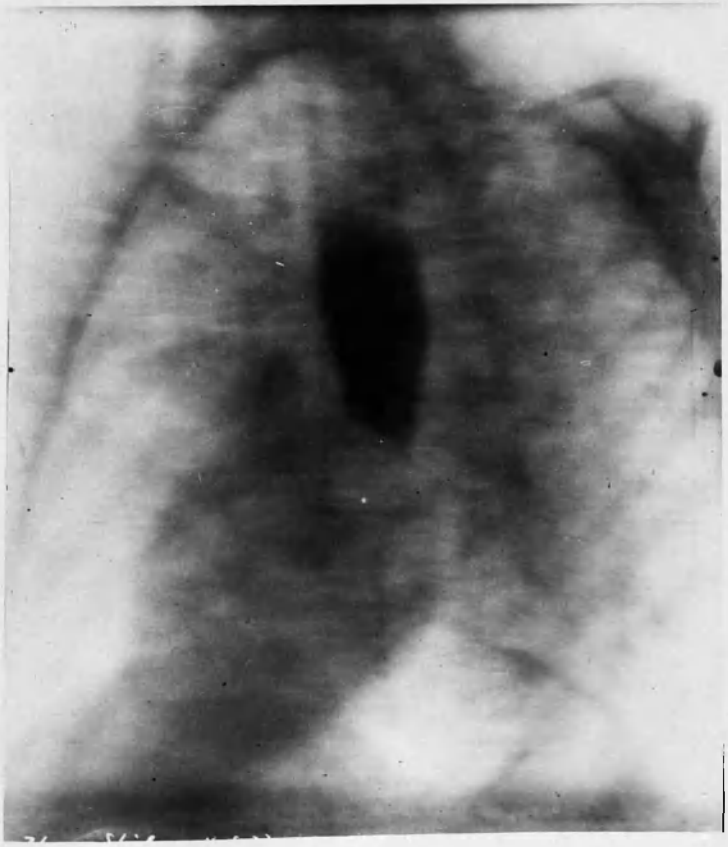


Fig. 27.