II. CONGENITAL NARROWING OF THE OESOPHAGUS.

It must have come under the notice of every observant person that there is a disparity in the ease with which different people swallow food. Remembering how certain individuals, who can swallow without thought the ordinary bolus, find it difficult or impossible to swallow a pill; and recognising that this inability is entirely a nervous disqualification, it is not to be wondered at if nervous instability be too readily accepted as the cause of all dysphagias where organic disease is not in evidence. But fuller consideration cannot fail to suggest that mere nervousness cannot be a satisfactory explanation of every non-pathological difficulty or delay in deglutition. An inconvenience to the individual, it is not fraught with danger to life, and perhaps on that account its significance has been overlooked and its importance underrated.

"Congenital narrowing of the oesophagus," writes Ballantyne,7 "with or without the oesophageal diverticulum, constitutes an anomaly which cannot be absolutely affirmed to be ante-natal in origin unless it be met with at birth, and since it does not prevent post-natal life (as does imperforation of the oesophagus), it is seldom that it is discovered at an early age." This is a somewhat misleading paragraph. Oesophageal diverticulum and congenital narrowing are two distinct anomalies in no way interdependent; and the recognition of constriction of the oesophagus in infancy depends not on its presence but on the degree of obstruction. The great majority of congenital narrowings cause no symptoms as long as the diet is fluid, and especially if the infant, breast-fed, has its natural and most suitable food. Should the constriction be extreme, even in infancy symptoms will follow, while with a less degree of constriction the existence of the anomaly will be noticeable only later in life, and dysphagia arise at less frequent intervals.

My attention was first seriously attracted to this condition
by the following case, which fortunately was an unusually severe one:

Case III.—Jessie was aged 12 years when she first came under my observation in August 1908, suffering from recurrent vomiting which had existed since infancy. She was the third child of a family of six. Her parents were well and normally developed, and the other children were well grown and each was healthy, though on a younger sister I had operated for a myeloma of the lower gum. Jessie was brought up on the breast only, till nine months of age. Almost immediately after birth she began to vomit occasionally, and this became more noticeable as she reached four or five months old. Her parents considered that it was on account of her difficulty in feeding that she did not grow as her brothers and sisters did. She walked at fifteen months. After the age of nine months she had skimmed milk added to her diet, and was given bread which had had boiling water poured over it, and the breast-feeding was gradually omitted. Any other diet brought on vomiting. With the vomiting she was never nauseated, and at meal times would take a little food, vomit it, and return at once for more. She had never at any time been able to take solid food. No pain ever preceded her vomiting nor accompanied her attempts to swallow. She could take liquid or thin soup, but milk was always her staple article of diet. Her parents recognised this and did not desire the child to be present at meals, but left instructions at a neighbouring dairy to give Jessie drinks of milk whenever she should ask for it, and in that way the child gained her sustenance. She was in hospital for a few days in 1900, and the condition, superficially investigated, was ascribed to gastric catarrh. Four years later, at the age of 9, she was again in a hospital and under observation for two months, and the illness ascribed to "habit vomiting." At this time an act, which in a younger or more sensitive child would have been one of peculiar brutality, was perpetrated on her. She was taken to another ward and shown a negro in bed, and told that if she vomited again she would be put to sleep with the "black man." The child prevented the threat being carried out by getting paper when she could—for she was allowed up in the ward—and picking out all the solids from her food, she wrapped them in the paper and surreptitiously disposed of them when she went to the lavatory. During this period of rest in hospital she gained 8 lb. in weight. At the age of 12 years she was a bright, healthy, active, and exceptionally intelligent child. She never vomited between meals. Any food not ingested came back exactly as it was taken, unaltered, not soured, but mixed with saliva, not containing mucus, and never containing blood. Nevertheless she was under-sized and under-developed, which perhaps made her intelligence and smartness the more striking. A photograph taken at this time standing alongside
The subject of the Congenital Narrowing of the Oesophagus has her sister, three years her junior, standing on her right.
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her sister, who was two years and eleven months her junior, conveys a better idea of her development than any description can. Under an anaesthetic the smallest size of oesophageal bougie was arrested at ten inches from the dental margin. A radiogram showed an elongated shadow in the region of the lower half of the oesophagus with a barium meal. If powdered biscuits were omitted from the mixture, and only barium and milk given, the fluid passed the stricture into the stomach, where it gathered as the usual rounded shadow in the left hypochondrium. Jessie was under my observation in the ward at that period during five months, and increased from 3 st. 5 lb. to 4 st. 2 lb., and for six months after her discharge she maintained this weight. For years I kept her under occasional observation. She passed through school creditably, and then became a worker in a mill, but at no time was she able to extend her diet beyond absolute fluids.

Were all cases as pronounced as this one, there would be little difficulty in persuading even the most prejudiced of the existence of congenital narrowing of the oesophagus. In accordance with all congenital defects, every degree is found. It requires little imagination to believe that had Jessie's malformation been a little more decided, it would have been incompatible with life; as it was, it seriously interfered with growth, and a condition which might be called "oesophageal infantilism" resulted.

In such a case as this, spasm would never be thought of; yet had it been less definite, spasm would certainly have been suggested, and by some most illogically insisted on. Its very severity puts it out of that category.

If cases of congenital narrowing of the oesophagus are rare, cases where the defect is noticed in infancy are rarer. The narrowness must be indeed extreme if regurgitation occurs before weaning brings about a change of diet and the ingestion of more solid food. Dunn has recorded the case of a boy aged 18 months in whom an obstruction could be felt by bougie 7½ inches from the teeth. Solid food was returned at once, but small quantities of fluid could be retained. A gastrostomy was performed, the stomach was found small, and by and by dilatation from below was attempted through a fresh gastrostomy opening. The child developed bronchopneumonia and gradually became emaciated and died, and post-mortem examination discovered a marked diminution in the size of the oesophagus at a point an inch above the diaphragm. Rogers records a very similar case of a male
child aged 22 months who was reported to have taken food well during the first ten months of life. An oesophageal tube could not be passed. The child lost weight rapidly, became bronchial, and died. Post-mortem examination "showed a fairly tight stricture about an inch from the lower end of the oesophagus."

Who could doubt that these two cases were congenital in origin? "Stricture" is the term used in each, but "stricture" is an objectionable word in that it conveys the idea of an inflammatory attack followed by pathological changes. Surely "congenital narrowing" would be less exceptionable. No philological objection could be raised to "stenosis" as descriptive of the condition, but I have avoided that term because it is apt to suggest a degree of contraction or closure which is only associated with the extremes of narrowing.

A very similar case to the one I have described is recorded by Whipham and Fagge of a female child of 4 years in whom vomiting had occurred from six months of age. Here, as in most other patients, the trouble began when weaning brings about a change in food. The child would eat greedily, and after three or four mouthfuls would say she was going to vomit, and without nausea would eject what she had swallowed. A bougie was arrested at the lower end of the oesophagus, while a No. 2 or 3 catheter passed easily into the stomach. Attempts to dilate the stricture caused perforation, and the child died three days later. Post mortem a fibrous stricture was found three-quarters of an inch below the bifurcation of the trachea. It was white, thick, and firm, and unlike the rest of the oesophagus. There was a perforation below the epiglottis, mediastinitis and right pneumothorax. I suggest that the changes in the oesophageal wall were the result of the interference.

"Narrowings are the most frequent and clinically the most important of the affections of the oesophagus," wrote Zenker in von Ziemssen's *Cyclopedia of Medicine* nearly half a century ago, and he pointed out that there are stenoses which from their anatomical structure (a perfectly healthy condition of the tissues and no trace of scars), as well as from their history, must be considered congenital, but which, in spite of considerable narrowing of the tube, might permit of a prolongation of life even to old age. These facts are well illustrated by the following case:
"Cardiospasm," Congenital Narrowing, etc.

Case IV.—In 1912, a few days before her death at the age of 82, I was asked to see Mrs S. because of her inability to swallow. In 1904 she had developed an ill-defined paresis, probably from atheromatous or senile changes in the cerebral blood-vessels. She was somewhat deaf and too ill to give any account of her symptoms. No tumour could be made out, nor were there any glandular enlargements. Her development was good and there was no emaciation. The smallest size of oesophageal bougie could not be passed through a stricture about the cricoid level. The provisional diagnosis was carcinoma. It was a surprise, therefore, on post-mortem examination, to find at the cricoid level a narrowing of the oesophagus through which only with difficulty could a small probe be passed. On this as a guide, the oesophagus was divided vertically, and examination then showed that all the coats were represented and all were normal. Each layer, mucous, sub-mucous, and muscular, was without any difference from the same layer in any other part of the oesophageal tube. Her relatives informed me that she had never been able to take any solid food, that she herself had declared that she had all her life had a narrow swallow, and had never been able to take food like other people. It was within their knowledge that she took only liquids, and that she had to swallow with slowness and care. Probably her death was due to senility, and only the senile weakness of the muscles and their enervation brought the lifelong dysphagia into unwarrantable prominence. As long as consciousness remained she was able to swallow liquids, but with the disturbance of consciousness she became unable to exercise the necessary care. This case affords other important points. There was no hypertrophy of adjacent parts, there was no dilatation, the anatomical parts were absolutely well developed and normal except that at the cricoid level for about an inch the calibre of the oesophagus was very greatly restricted.

This case also shows that congenital narrowing is not limited to the lower end of the oesophagus. Indeed, though, as would be expected, more common at either extremity, no part of the tube is exempt from this congenital defect.

Sir Everard Home records the case of a lady of 46 who came under his observation. She had "from her earliest remembrance had a narrow swallow," which had given more trouble during the preceding two years. She had "attacks threatening suffocation," in one of which she died. The orthopnoea was evidently in association with the extensive disease of the right lung which was found on post-mortem. The oesophagus immediately behind the cricoid was contracted. There was no thickening, and the stricture, Home says, consisted
of a fold of the internal membrane only. The specimen is in the museum of the Royal College of Surgeons of England (No. 2296), and it is noted in the descriptive catalogue that “no trace of stricture can now be seen.” It is curious to see the specimen after so many years (Home’s book was published in 1803) still blackened by nitrate of silver, the application of which Home claimed to be the successful treatment of such strictures.

There are other specimens of narrowing of the oesophagus in the Royal College of Surgeons’ Museum, and a few I have examined in other London museums, but there is a lack of evidence or history that would prove their congenital origin, but at the same time there is also a lack of proof of later acquisition. Another difficulty than want of previous history in collecting cases on which to base a justifiable conclusion is the want of sufficiently prolonged observation. Butlin, after referring to the rarity of chronic obstruction to swallowing in young people, tells of a boy of 11, whose mother stated “that never in his life had he been able to swallow solid food.” He was well nourished and well developed. Under an anaesthetic Butlin passed a bougie. “About the middle of the oesophagus,” he writes, “it seemed to encounter some hindrance, and overcoming that hindrance by a little pressure I passed it into the stomach.” While stupid with the anaesthetic the boy swallowed a lump of bread, but refused to attempt such a thing when he regained his senses. Butlin believed he had broken down a congenital diaphragm in the oesophagus. In the same paper he records a second case—that of a girl of 15, emaciated and under-developed, with a history that she had never been able to swallow solid food, and even fluids she swallowed with difficulty. An attempt to get in oesophageal tubes had to be abandoned, but the attempt seemed to have done good, for gradually thereafter the power of swallowing returned until she was able “to take food like any other child.” Butlin again believed that he had broken down a congenital web, and that the child was practically cured. In a short reference to congenital stenosis of the oesophagus, Kirmisson quotes a case on the authority of Mayer of New York, of a female aged 9 years, who from birth frequently regurgitated milk along with quantities of stringy mucus. The vomiting was unaccompanied by any abnormal contractions. “The sound revealed a marked stenosis in the lower part of the oesophagus.” After three months of progressive dilatation the
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child could take solid food, and at the end of a year was “restored to normal health.”

Butlin's idea of a congenital web was of course not original. It was an old suggestion, promulgated before the ontogenesis of the oesophagus was known, and doubtless stimulated by the clinical observation of the less varieties of imperforate anal canal in which the meconium shows through a safely incised membrane. In more modern times the occurrence of narrowings or diaphragms have been described as being present in other parts of the alimentary canal, but in association with foetal structures or at the junction of morphologically separate units, and it did not need the title of Home's monograph, "Practical Observations on the Treatment of Strictures in the Urethra and in the Oesophagus," to suggest a relationship between these two canals. If there were any similarity, however, between web-obstruction in the oesophagus and in the urethra, a paper by Young, Frontz, and Baldwin would dispel the illusion. In their paper entitled "Congenital Obstruction of the Posterior Urethra," they describe in their "Case IV." a thin, fibrous-looking membrane at the apex of the prostate, attached to the entire circumference and forming pockets with their concavity towards the bladder, and this they allege bringing about hypertrophy of the bladder, dilatation of the ureters, and hydronephrosis. Were there any analogy one would expect some proximal hypertrophy in oesophageal web-obstruction, whereas such overgrowth is unknown either in web-obstruction or congenital narrowing.

The analogy between the oesophagus and the pylorus is so erroneous and misleading that the advisability of even mentioning the two together seems almost unjustifiable, but the deduction drawn from urethral obstruction may be amplified by reference to a communication on congenital pyloric stenosis by Cautley and Dent. They figure in the obstructed pylorus a longitudinal fold of the mucosa. To this fold the writers attribute a causative importance which the experience of other observers has not confirmed, but it is not illogical to infer that were that longitudinal fold the cause of the obstruction, a web-fold in the oesophagus ought to be associated with equal hypertrophy to the pylorus; and further to induce, that if the longitudinal fold were secondary to the pyloric obstruction, a web-fold in the oesophagus might have some causal relation with that congenital narrowing which is so frequently
noted as being present along with the so-called “congenital web.”

About the third week of development of the embryo the human oesophagus is merely an annular constriction between the pharynx and the stomach, and it is only with the differentiation of the neck that elongation takes place. Were, as a result of maldevelopment, some of this constriction to remain, it is conceivable that something of the nature of a diaphragm might remain (though an areal and not a horizontal linear constriction is much more conceivable), but I do not think that such a diaphragm would consist of internal coat only. If the “web” caused appreciable obstruction, undoubtedly the oesophageal muscle proximally would alter its normal condition by becoming hypertrophied.

There is a Hunterian specimen in the Royal College of Surgeons’ Museum (No. 2294) in London which is described as “part of the pharynx with larynx. Opposite the lower margin of the cricoid cartilage there is a projecting annular fold of mucous membrane about a line in depth, narrowing the passage into the oesophagus. The adjacent parts appear healthy, and the fold is composed of healthy mucous membrane. The canal around the fold is slightly constricted.” The origin of the fold may be as suggested, but the information about the specimen and its origin is insufficient to justify speculation. It is to be noted that the fold is at the entrance to the oesophagus, and is not associated with either inflammatory changes or hypertrophy.

The specimen exhibited next to this one in the same museum is again from the collection of Sir E. Home (No. 2295). It is from a child, and “at the level of the lower margin of the cricoid cartilage, the commencement of the oesophagus is reduced to less than half its natural diameter by a sudden contraction of its walls. The tissues at and around the stricture are not visibly altered, and the canal above and below it is healthy.”*

Home mentions that of cases of oesophageal dysphagia which came under his notice: four were in ladies of delicate constitution who had noticed a difficulty in swallowing before 17 years of age, and that this difficulty had increased at the age of 40. That is not to be wondered at, for as the resiliency of youth passes off the oesophagus, in common with other passages, becomes less distensile and the general musculature less active.

Under the title “Simple Stenosis of the Gullet,” Sir Morell Mackenzie includes consideration of cases which present

* My italics.
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"abnormal narrowness of a limited portion of the oesophagus without any morbid change in any of its component tissues at the seat of stricture.” To the few references he has collected he adds no case of his own, so that his conclusions, drawn from information and not from observation, lose considerably in value. His definition is excellent, but he lacks consistency when he includes under “simple stenoses” cases of partial paralysis and dilatation. All the same, it is obvious that this authority recognised the existence of true congenital narrowing of the oesophagus.

In a paper dealing with carcinoma of the post-cricoid region and upper end of the oesophagus, Logan Turner\(^1\) states that out of thirteen men and eighty-one women suffering from this condition, four women “stated that as long as they could remember they were obliged to eat slowly, explaining the fact on the ground that they had ‘a narrow throat.’” If these were cases of congenital narrowing of the less degree, almost 4 per cent. seems a very high ratio amongst cancer; but it must be remembered that carcinoma tends to occur at points of irritation as well as at the junction of heteromorphological areas, and certainly deglutition will cause more irritation at a point of congenital narrowing than in a normally distensible tube. Indeed, in this relation Logan Turner writes: “It is now the experience of many laryngologists that a considerable proportion of the women who suffer for a number of years from spasmotic difficulty in swallowing, eventually develop carcinoma in the laryngeal part of the pharynx.” I would take exception to the term “spasmotic difficulty” used here by Logan Turner. What these patients really complain of is “intermittent difficulty,” and the laryngologist, without realising that he is approving a theory, dubs it “spasmotic.”

The following case illustrates one of the minor degrees of congenital narrowing:

**Case V.—** David W., a draper, aged 68, stated in 1912 that he had “naturally a narrow gullet and had been sensible of that all his days.” He could not recall any particular occasion when his usual care in swallowing had been followed by any untoward event until about 1897, when, while eating a tough rabbit, a portion had stuck in his gullet. A medical man was sent for, but before he arrived relief was obtained by the bolus passing into the stomach. A stomach tube, however, was passed, more to justify the doctor’s visit than to relieve the patient’s necessity. Some years later the inability to swallow easily
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caused him to consult Dr Malcolm, who regarded the condition as functional. In 1907 he was eating a variety of stone fruit known as "Victoria Plum," and turning his head sharply to make a remark he swallowed a stone. He felt the stone "somewhat tight in going down." As he had finished eating he did not swallow anything more that night, but went to bed as usual, and next morning he rose to take his breakfast. He found, however, that he could not swallow anything, nor was he able to swallow at all during the entire day, and by night he was prostrate with exhaustion. During the subsequent few weeks his food consisted entirely of milk and cod-liver oil, poached eggs, and pease-meal brose. In January 1908 he was examined successively by three surgeons. Two of these attempted to pass an oesophageal bougie, but each found a distinct obstruction preventing its entrance to the stomach. He was told that he had cancer of the gullet, and that his emaciation and exhaustion made him unsuitable for surgical interference. The obstruction continued during one year and ten months, when he rose one morning to find the obstruction gone. He began to eat, and was astonished to find he could swallow anything, though of course he resumed the usual care which had characterised his eating during his entire lifetime. In 1914 he continued well, though he had to be careful with dry food as he had always had to be.

The lifelong necessity for care in swallowing seems sufficient proof of congenital origin in this case. The absence of discomfort when the blocking occurred surely eliminates spasm, and to my thinking the duration of the obstruction during sleep and waking and its sudden relief is incompatible with muscular contraction.

Case VI.—Before the days of radiography a child, aged 3 years, was admitted to hospital with vague symptoms of chest trouble. He objected to lying down and had a slight rise of temperature, but took the fluid and semi-solid food which was given him quite well and without apparent difficulty or delay in swallowing. After a residence of some days pleurisy became obvious and the child died. On post-mortem examination the oesophagus was found perforated by an impacted halfpenny, which had produced inflammation in the mediastinum. Such cases emphasise, what is the fact, that spasm may not be a prominent accompaniment to foreign bodies impacted in the oesophagus. It is also noteworthy that the diminution of the oesophageal calibre was not accompanied by regurgitation or difficulty in swallowing fluid food.

Sir Sinclair Thomson records the successful removal per oram of a tooth-plate which had been impacted in the
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oesophagus during two and a half years, which rendered the patient unable to swallow solid food. He quotes a case recorded by Guisez of a ten-centimetre piece having lain in the oesophagus for four years. These, however, are nothing to the case referred to by Tilley,20 in which "Brandon Kyle, Philadelphia, removed a denture after seventeen years' sojourn" in the oesophagus! Finally, in this connection, Kahler21 records the case of a man sent to hospital as a case of carcinoma of the oesophagus of four weeks' duration, in which the oesophagoscope demonstrated the impaction of an artificial denture. In these days of radiographic examination such occurrences become less likely.

It would not be right to dismiss the subject of congenital narrowing of the oesophagus without a word about treatment. Recognition of the condition is the essential and is the treatment. It must never be forgotten that in all forms we are dealing with a congenital defect, and congenital defects are notoriously difficult and unsatisfactory for operation. The anatomical structures are fully developed as far as the embryonic formative material allows, and no mechanical procedure can add to its amount. As regards children, the passage of a bougie, the brutality of a threat, the mental dread of asphyxiation by an anaesthetic, may each or all suggest to the child to have recourse to subterfuge—the refuge of the physically inferior. The only treatment is appreciation of this congenital deformity and the modification of diet or eating throughout life to suit the circumstances. This appreciation will never be obtained until text-books recognise the entity congenital narrowing of the oesophagus is, its syndrome, and its clinical importance.

(To be continued.)