HYPERTROPHIC PYLORIC STENOSIS IN THE ADULT

SAMUEL SKAPINKER, M.B., B.CH., F.R.C.S. (EDIN.), and R. GLYN THOMAS, M.B., B.CH., D.M.R.D.,
Johannesburg

This condition has been known since ancient times, and credit for the first description is accorded by Kellet to Fabricus Hildanus for his 'Observanti Singularis de Obstructi Pylori'. Other early reports were by Blair in 1717, Webber in 1758, and Armstrong in 1771.

Jean Cruveilhier[4] reported the case of a 72-year-old female with hypertrophic pyloric stenosis in 1835 and Maier reported 31 cases in 1885. Tiegler in 1893 collected 23 cases from the literature.

Today most clinicians are aware of infantile hypertrophic pyloric stenosis, but few of the occurrence of hypertrophic pyloric stenosis in the adult.

The Anatomy of the Pylorus

The anatomy, physiology and pathology of the pyloric region were correlated by Torgersen.[18] His investigations showed that the pyloric canal in man contains a specialized muscle fibre forming the pyloric ring. This muscle consists of sphincter loops enclosing interpositioned circular fibres. The distal sphincter loop represents the thickened bundle of circular muscle fibres corresponding to the gastric portion of the pyloric sphincter. One proximal sphincter loop embraces the proximal end of the pyloric canal obliquely and forms the intermediate sphincter. The fibres of the pyloric and intermediate sphincters converge on the lesser curvature forming a muscle prominence — the muscle torus.

Classification

Skoryna, Dolan and Gray[15] proposed the following classification:

Primary pyloric hypertrophy
1. Focal form
2. Diffuse form with proximal lesions
3. Diffuse form without proximal gastric lesions

Secondary pyloric hypertrophy
1. Associated with distal obstructive lesions

Incidence

This is difficult to ascertain since many of these cases are unreported and many have such mild symptoms that they are not diagnosed.

The largest series reported is by Kirklin and Harris[7] (1933) who described 59 cases of hypertrophic pyloric stenosis occurring in patients at the Mayo Clinic in a 5-year period. North and Johnson[11] collected 59 cases and added 5 of their own. Craver[2] reported 11 cases confirmed at operation at the New York Hospital, Cornell Medical Centre, over a 20-year period. The most recent review was by Knight,[8] who described 7 cases in 1961.

The age of the patients varies with different series and, although the condition has been described in 14-year-old children, it has also been described in an 85-year-old patient.[11]

Men have been more commonly affected than women. This, interestingly, corresponds to the sex incidence in congenital pyloric stenosis of infants.

Aetiology

It has been suggested that both the adult and infant forms represent a persistence of the relatively thick pylorus normally present from the fifth to the sixth month of embryological development. Crohn[3] felt that it was a persistence of the infantile type into adult life, and fewer cases will be found now that infants are being treated more vigorously. Woo-Ming[20] describes a family of a father of 49 with adult pyloric stenosis and a son of 13 who had a congenital pyloric stenosis, and also relates that Fenwick[5] reported a man with pyloric stenosis who had 2 sons with congenital pyloric stenosis. Spasm of the pylorus has been suggested as a cause, but this has been rejected — mainly because in no other part of the body does a smooth-muscle sphincter undergo hypertrophy from spasm. Another factor is that although pylorospasm is common, pyloric hypertrophy is rare.

Pathology

The gross appearance in adults is the same as that found in infants. A fusiform mass is present, which is thickest at the duodenal end and gradually thins into the antrum as it passes proximally. The outstanding feature is the hypertrophy of the circular muscle layer of the pylorus.

There are 2 main pathological differences between pyloric stenosis in the infant and the adult. These are:

1. The hypertrophy involves the whole circumference of the pylorus in the infant, but in the adult it may involve a localized segment.

2. Raia *et al.*,[14] in studying the pathogenesis of pyloric stenosis of the newborn and comparing it with the adult form, found that there was a marked difference in the mesenteric plexus. Where there is degeneration of the plexus in the newborn, this is slight and reversible, while in the adult the degeneration is so marked that there is no hope of reversibility. This parallels Hurst's theory of achalasia.

Histological section reveals marked hyperplasia of the muscles.

Clinical Picture

The symptoms may commence and persist from infancy or may appear in adult life. In 70% of the collected cases the symptoms commenced between 31 and 60 years of age.

Bockus[1] divides the symptom-complex into 3 groups:

(*a*) Signs and symptoms from childhood.
(*b*) Those with atypical ulcer symptoms of long duration beginning in adult life.
(*c*) Those who quickly develop signs and symptoms of pyloric obstruction in middle or late life.

The patient may complain of pain, frequent eructations, epigastric heaviness, postprandial pain, and night regurgitation of acid material. In those patients with associated gastric ulcer the symptoms of the ulcer may overshadow the symptoms of the stenosis. The gastric analysis is of no assistance since it may vary from hyperchlorhydria to achlorhydria. Our patient presented with achlorhydria.

Weight loss is a frequent finding. Although found commonly in the child, a mass is seldom felt in the adult.

The incidence and the relative significance of associated gastric lesions are controversial. Only one of Knight's 7 patients had a gastric ulcer. Texter *et al.*[17] reported a study of 55 patients with ulcers in the pyloric canal. In this group the main symptoms appeared to be a typical pain, nausea, and vomiting. Weight loss and low gastric acid was not uncommon. In fact, the symptoms were remarkably similar to hypertrophic adult pyloric stenosis, except that in the latter severe pain is absent.

Radiological Features

The pyloric canal is narrowed and elongated from its normal half-to-one-centimetre length up to 4 cm. or more. The transition from narrowing to normal width is abrupt at the gastric end, and smooth, rounded bulging of hypertrophied muscle into the gastric antrum is sometimes seen. The muscular walls can contract to obliterate the lumen, but do not relax. The narrowed pyloric segment is unaltered by medication or manipulation.[10] The symmetry of the stenosis depends upon the particular portions of muscle involved by the hypertrophy. The canal usually lies closer to the lesser curve aspect of the antrum than the greater curvature.[7]

The hypertrophied pyloric sphincter indents the duodenal cap to give a mushroom-shaped pressure deformity of the base of the duodenal bulb. Twining[19] has described this as resembling an os uteri at operation.

Gastric hold-up of barium is frequently present, but is usually less than with a stenosing ulcer or carcinoma, amounting to about 25% at 6 hours.[6] Hyperperistalsis is frequently present, the waves being deep but slow. There may be a niche between the pyloric and antral components of the muscle hypertrophy with a retained barium fleck mimicking an ulcer. However, the barium flecks tend to be inconstant. The mucosa at the entrance to the narrowed segment is frequently contracted, with retained barium pockets, but the folds become narrower, finer, and more longitudinal within the narrowed segment.

Some authors describe a 'palpable resistance' in the region of the pylorus.[13]

Differential Diagnosis

Pre-operative diagnosis may be difficult. In our patient the dyspepsia, weight loss, achlorhydria and X-ray findings strongly suggested a carcinoma of the stomach. In retrospect, had we thought of the condition, we might have made the diagnosis.

The radiological differential diagnosis of the deformity of the pyloric canal includes simple antral spasm, stenosing pyloric ulcer, and scirrhous stenosing carcinoma. Confusion with an encircling adhesion about the pylorus is mentioned by Pendergrass.[12]

Rare conditions such as antral gastric syphilis, lymphoma infiltration, or an eosinophilic granulomatous mass may be difficult to distinguish from pyloric stenosis, clinically and radiologically.

The mushroom-shaped indentation of the duodenal cap which, after the appearance of the pyloric canal, is the most distinctive feature of the condition, can be imitated by the transpyloric prolapse of gastric mucosa and by variations in radiographic projection.

Treatment

The treatment of hypertrophic pyloric stenosis in the adult is surgical, especially if the patient presents with symptoms or if malignancy is suspected. At operation, as in our patient, the condition is easily recognized and treated as such. The methods of treatment described have varied from simple dilatation to a Ramstedt type of pyloroplasty and other forms of pyloroplasty. Pyloroplasty has been used successfully, but an adequate biopsy cannot be obtained and a malignant growth may be overlooked. A gastrectomy should be performed if the patient is suf-

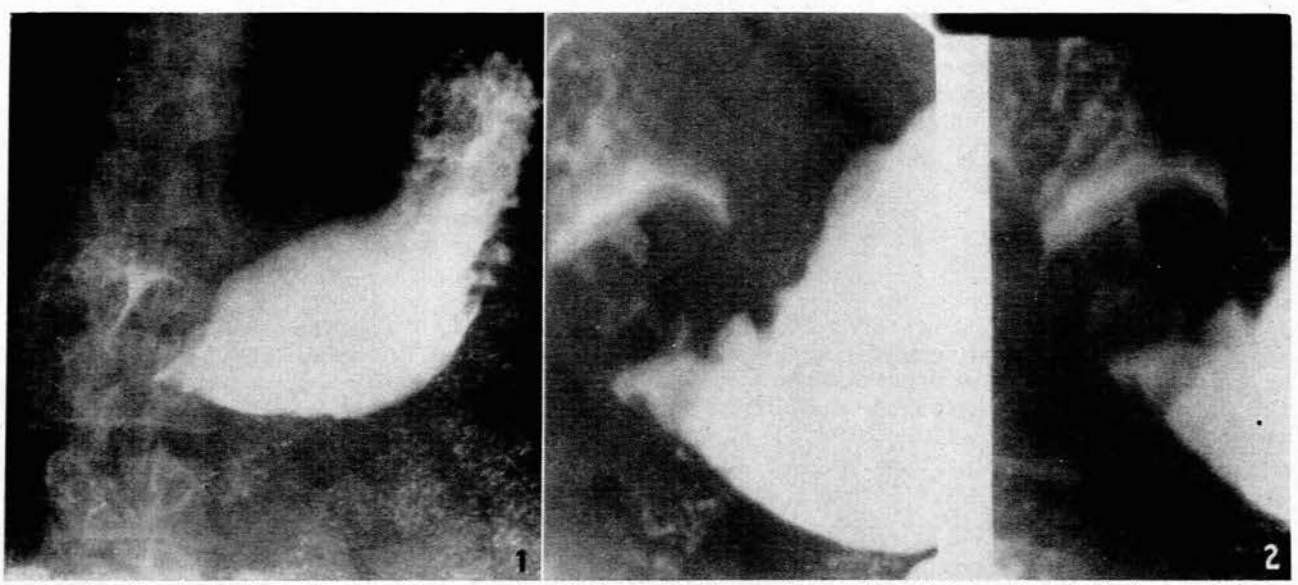

Fig. 1. This shows the pyloric canal narrowed and elongated with mushroom-shaped deformity of the base of the duodenal cap.
Fig. 2. From serial-strip radiographs; the barium fleck within the narrowed pyloric lumen is demonstrated.

ficiently fit. The results of simple gastroenterostomy have been unsatisfactory and the symptoms have not been relieved.[9,16] Knight[8] puts gastrojejunostomy in the category of 'not recommended'. In most series the best results were obtained by gastrectomy.

Case Report

Mr. P.R., aged 51, a White male, was referred to us by Dr. I. Chavkin. His history was mainly of belching air, and this was more marked on holiday. The patient ascribed this to the change of diet. He had marked acid regurgitation which lasted for about one minute. At the same time he became bilious. These symptoms commenced 3 years ago. For the past few months he had marked loss of weight and general loss of appetite. The remainder of his history was non-contributory. He was examined by Dr. D. Lurie, physician, and had a normal electrocardiograph.

Examination. The patient was well covered and had no tenderness. Gastric analysis revealed a resting achlorhydria with no response to alcohol or histamine. Excess mucus was present. The blood count was essentially normal. There was no anaemia.

Radiological examination. Barium-meal examination showed marked narrowing of the prepyloric antrum of the stomach with mushroom-shaped indentation of the base of the duodenal cap and a niche in the narrowed area, which was interpreted as an ulcer crater (Figs. 1 and 2). Despite vigorous peristalsis and the use of 'buscopan' intravenously, gastric emptying was very slow and there was an hour's delay before the duodenum filled. A very large 2-hour gastric residue of barium was present.

Treatment. In view of the achlorhydria, X-ray findings, and weight loss it was decided that a laparotomy and gastrectomy should be performed.

On 24 March 1961 an exploration through a right paramedian incision demonstrated a hard indurated pylorus, and a Hoffmeister-Polya gastrectomy was performed removing about three-quarters of the stomach. Convalescence was uneventful.

Dr. John Gluckman, pathologist, reported:

'Sections were prepared from various parts of the portion of stomach with pylorus. Microscopic examination revealed a moderate degree of gastritis with associated oedema. The muscle bundles were widely separated by that process. There was some hyperplasia of the lymphoid follicles. This was entirely non-specific. The essential pathological changes here appeared to be very marked myohyperplasia with oedema.'

Follow-up 7 months later was extremely satisfactory. The patient was asymptomatic and eating well. He had regained all his lost weight.

SUMMARY

1. Hypertrophic pyloric stenosis of the adult is discussed from the aetiological, anatomical, and radiological viewpoints.

2. The surgical treatment advocated is gastrectomy.

3. A case report is presented.

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