

1931

## Congenital hypertrophic stenosis of the pylorus

Marvin Nerseth  
*University of Nebraska Medical Center*

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SENIOR THESIS

CONGENITAL HYPERTROPHIC STENOSIS OF THE PYLORUS

UNIVERSITY OF NEBRASKA

1931

MARVIN NERSETH

CONGENITAL HYPERTROPHIC PYLORIC STENOSIS

INTRODUCTION

Definition

Of the various terms used at one time or another to define this condition, congenital hypertrophic pyloric stenosis is perhaps the most descriptive and most nearly in accord with the theories of present day observers. The term "congenital," indicates the possible time of origin of the lesion, and the term "hypertrophic" suffices to suggest the pathological picture.

Richter (1) in Abt's Pediatrics, gives a rather complete definition of the condition. "Congenital hypertrophic stenosis of the pylorus is a mechanical obstruction of the alimentary tract, due to a simple hypertrophy and hyperplasia of the circular muscle fibers of the pylorus, of congenital origin, manifesting itself soon after birth by all of the usual symptoms of intestinal obstruction, modified by the location of the lesion".

History

The disease is by no means a new one, but until thirty years ago it was regarded as a pathological curiosity, diagnosed only after death and invariably fatal. Today, while it does not commonly occur, it presents a symptomatology so definite, and responds so readily to early treatment, that a knowledge of it's diagnosis and treatment is quite essential.

Caulfield (2) attributes the earliest description of the disease to Patrich Blair who reported a case in 1717 to the Royal Society of London. This report gives a definite clinical history of the condition as it is now known, and is substantiated by autopsy findings.

The first essay on the subject recorded in American literature is accredited by Foote (3) to George Armstrong. Armstrong, in 1771, describes a case in an infant three weeks of age. The clinical history is very accurate and post-mortem examination reveals a mass in the pylorus.

In 1788, Beardsley (3) reported a case to the Medical Society of New Haven County, entitled, "A case of Scirrhus in the pylorus of an infant." From this time until 1841 there are no other cases on record. In 1841 Williamson (1) of London, under the title, "A case of Scirrhus of the stomach, probably congenital." describes the condition in a male infant. The following year Siemon-Dawosky (1) published an account of the first case described in Germany, and called attention for the first time to the projectile character of the vomiting.

From 1841 until 1888 the condition received very little consideration. Hirschsprung (4) in 1888 aroused an interest in the study of the pathologic anatomy of hypertrophic pyloric stenosis. He describes the symptoms and gross pathology of the

pylorus in several instances. From these observations he formed the theory that the stenosis was due to a simple hypertrophy and hyperplasia of the circular muscular layer in the pylorus.

In a review of the literature made by Cautley (5) in 1902, he finds that until that date, fifty cases had been recorded. It was at this time that he reported a case in a seven months foetus, which suggested the possibility of the condition having a congenital origin.

Since 1902, the condition has been given proper attention in medical literature and it's diagnosis and treatment has been well established.

#### ETIOLOGY

The exact etiology of congenital hypertrophic pyloric stenosis, is as yet quite undetermined. Various theories have been advanced by different authors during the past thirty five years.

The condition has no racial prevalence, and very slight, if any, geographic limitations, Eastern Europe being the only portion of the world in which it is seldom described. It occurs more frequently in male than in female infants, the ratio being approximately four to one.

Dounes and Bolling (6) state that it occurs in one out of every two hundred male infants. A large proportion of the cases

occur in first born male infants, and the frequency is greater among infants which are breast fed. In a series of four hundred fifty four cases studied by Bolling (27) the age of onset was during the fourth week. Steen 22 states that in fifty percent of his cases the age of onset was during the third week.

There appears to be some familial tendency connected with the condition. This cannot, as yet, be attributed to either hereditary or environmental influences. Numerous cases have occurred in the same family, and in twins. Two cases have occurred in successive generations.

Two cases occurring in the same family are reported by Moore (7). In both instances the victims were male infants.

Bilderbach (8) cites the condition occurring in premature twins. Both infants were operated upon by the Fredet Rammstedt method with recovery.

The occurrence in successive generations is described by Caulfield (9). This is the only instance of this nature found in the literature. The first case was not under Doctor Caulfield's observation and occurred before surgical treatment of the condition was popular. The history and symptoms were typical of the condition, and the patient responded to medical care. The next case occurred in the first born daughter of the previous patient and was under Doctor Caulfield's care. Typical symptoms began at

the age of three weeks. Operation was performed and a tumor was found, in the pylorus, which was diagnosed as hypertrophic stenosis.

Hirschsprung (4) in 1888 advanced the theory attributing the condition to a primary hypertrophy of the circular muscle layer of the pylorus. This produced a reduction in size of the pyloric lumen resulting in obstruction. It is at the present, generally believed that the hypertrophy in the given case is insufficient to produce obstruction, but that an accompanying spasm must be present. There is much controversy between authors as to which is present first. There is no evidence to show that spasm causes hypertrophy, it may occur at any age, but it does not produce the condition of hypertrophy which is in general, peculiar to the first few weeks in life.

In 1902 Cautley discovered a typical hypertrophy of the pylorus in a seven months foetus. The tumor had all of the characteristics of one removed from an infant having the disease. This has given a congenital aspect to the condition indicating that the hypertrophy precedes the spasm and that it develops in utero.

In 1922 Haas (10) stated that the condition is simply a dominant feature of a general state of hypertonicity. Hypertonicity of all skeletal and smooth muscle exists. The gastrointestinal tract shows increased activity simulating spasm and is due to an overactivity of the vagus nerve, known as vagotonia.

Hypertrophy of the pyloric sphincter results from overwork, and continued spasm brings about stenosis. Haas bases his theory on the fact that atropine relieves the obstructive symptoms by direct action on the vagus nerve.

Cushny (11) has shown that the vagus nerve continues to exert its normal influence on the intestines after the therapeutic administration of atropine and that therapeutic doses simply inhibit abnormal peristaltic movements. Haas's theory therefore, is not conclusive. Only a small percentage of cases are permanently relieved by atropine.

Brodie (12) and his co-workers have experimentally produced hypertrophic pyloric stenosis in rats with results nearly identical to the condition in humans. The experiments consisted of feeding pregnant rats on a diet with a deficiency of the anti-neuritic vitamin (B). A control group was run at the same time and given a normal diet. In twenty three of the offspring from the rats fed on the diet deficient in vitamin B, ten developed true hypertrophic stenosis of the pylorus. Of the control group, all offspring were normal. The obstruction was much more common in young rats of the second generation and seven eighths of the cases occurred in males. The vagus nerve in all of the cases showed myelin degeneration.



From these experiments it is suggested that the condition <sup>may be</sup> due to a vitamin deficiency resulting in a nerve involvement. The vagus being motor to the pyloric sphincter, irritation in utero from myelin degeneration is capable of producing contraction of the muscle. The prolonged contraction resulting in hypertrophy.

The authors suggest that dessicated yeast be added to the mother's diet as a prophylactic measure.

This is an interesting experiment, if the disease can be proven to fall in the group of vitamin deficiencies, much may be accomplished along prophylactic lines.

Pirie (13) considers hyperadrenalism to be the cause of pyloric hypertrophy by creating a pancreatic insufficiency which accentuates pyloric closure.

The pylorus and medulla of the suprarenal glands become differentiated at about the third month of intra-uterine life. A hypersecretion of adrenalin produces a contraction of smooth muscle. This being the case, there would be plenty of time for spasm induced by any excessive supra-renal secretion to bring about hypertrophy. He makes no claim that adrenalin has any selective action on the pylorus alone, but that the peculiar anatomical and secretory relations existing between the stomach, pylorus and duodenum play an important part. The supra-renal gland before birth is larger than the normal kidney, therefore an early hypersecretion would be quite possible. The reason that true hypertrophic stenosis is rather infrequent, is due

due to the fact that the degree of hyperadrenalism is not constant.

Pirie believes that the hypertrophy produced by hyperadrenalism is in itself, insufficient to produce complete obstruction. He claims that the obstruction develops when spasm is added, and that this spasm is brought about by the peculiar secretory and anatomical relation existing between the pylorus stomach and duodenum. The opening of the pyloric sphincter is brought about by the presence of the acid chyme in the stomach, and that after the acid chyme reaches the duodenum, the valve closes. The passage of this into the duodenum stimulates the formation of secretin, which is carried by the blood stream to the pancreas. The pancreatic secretion into the duodenum is stimulated by secretin, and its action in the acid chyme is that of neutralization. Therefore, due to the partial obstruction in the pylorus already present from the previous hypertrophy, there is a diminishing in amount of acid chyme entering the duodenum. This results in a lessened amount of secretin produced and a corresponding diminution of pancreatic secretion. Hence the acid chyme is not completely neutralized, causing a prolonged closure of the pyloric sphincter.

In summary of Piries theory, the pyloric hypertrophy is caused by hyperadrenalism, and the added spasm causing obstruction results from a pancreatic insufficiency.

This theory is not intirely conclusive. The hypertrophy may be due to hyperadrenalism, but physiologists disagree on the theory of acid control of the pylorus. Thomal and Wheelon (14) have shown that by changing the contents of the stomach and duodenum from an acid to an alkali media and visa versa, pyloric closure or relaxation was not affected. They found that there may be a complete achlorhydria, and yet the movements and emptying of the stomach presented nothing abnormal.

Some interesting experiments and observations regarding the allergic nature of pyloric stenosis have been carried out by Cohen and Breitbart (15). They contend that the organic change in the pylorus is usually insufficient to cause complete obstruction. Spasm of the muscles and edema of the mucosa and sub-mucosa act as adjuvants and complete it. Infants take food and gain in weight for a few days and then suddenly develop symptoms of pyloric obstruction. Something of an acute nature must have set in and completed the already existing partial obstruction.

It has been found by the students of anaphylaxis that the characteristic picture in anaphylactic shock, consists in smooth muscle spasm, edema and exudation. The intestines of a rabbit were observed in situ during an anaphylactic shock, following the administration of a dose of horse serum. Spasm of the smooth

muscle and actual peristaltic rushes were noted.

This led to an investigation of a series of cases of pyloric obstruction from an allergic point of view. They were studied as to (a) the presence of other allergic reactions, (b) the demonstration of specific skin reactions to definite allergins and their transfer by the method of Prausnitz and Kustner, and (c) the therapeutic testing of dietary change, based on the evidence obtained by the immunologic method.

Accurate immunologic studies were made on six infants, four at the time of obstructive symptoms and two later in life. Of the four studied during the active symptom period, skin reactions to egg, milk or cereals was obtained, and three successful transfers by the method of Prausnitz and Kustner were made. In the two cases studied later, both infants developed a marked eczema, one was found positive to egg and the other to milk.

Three typical cases of congenital hypertrophic stenosis are reported. In each instance there is a familial history of allergy, and in one a familial history of congenital hypertrophic stenosis. Positive sensitization tests were obtained in all three cases. By dietary change, that is ~~the~~ elimination of the food to which they were sensitive, and by medical treatment, definite improvement resulted.

The organic pyloric obstruction is believed to be due to sensitization in utero, early in pre-natal life. Repeated shocks at this time resulting in a secondary hypertrophy of the pylorus. The child develops normally for a time after birth, and then receives a dose of the specific allergin which results in a mild anaphylactic shock, manifested in spasm of the pyloric sphincter and edema of the adjacent tissues. This, together with the preexisting hypertrophy, is sufficient to cause complete obstruction.

This work of Cohen and Breibart has introduced the prophylactic and therapeutic treatment of congenital hypertrophy of the pylorus into a new field of medicine, namely that of allergy. The results and observations however, have been based on an insufficient number of cases to draw any definite conclusions. It is interesting to note the incidence of the condition with that of an allergic history and individual allergic manifestations. Their work is worthy of further study and experimentation, and the results may introduce a specific preventive and non-operative treatment.

## ANATOMY

Cunningham: (16) The normal pyloric canal in an infant presents itself as a cylinder approximately 1.5 centimeters in length. It is generally contracted and its lumen obliterated by closely packed longitudinal folds of mucous membrane. The pylorus protrudes into the duodenum and gives the impression of <sup>a</sup> miniature vaginal portion of the cervix uteri.

The pyloric orifice is located at the extremity of the pyloric end of the stomach, and its position is indicated upon the surface of the stomach by a slight annular constriction, which is most marked at the curvature termed the pyloric ring. The constriction marks the junction of stomach and duodenum, and there the various coats of these portions meet with one another.

The peritoneal covering of the stomach is continued onwards to the first part of the duodenum. The longitudinal muscle fibers radiate from the esophagus in all directions, ~~ending~~, however, ~~before~~ they reach the pyloric region. A new set of longitudinal fibers takes origin in the body of the stomach and these become more numerous as they are continued over the pyloric canal, forming a distinct longitudinal coat. Comparatively few of these fibers pass over to become continuous with the corresponding fibers of the muscular coat of the duodenum. The deeper longitudinal fibers

leave the surface and penetrate the substance of the pyloric sphincteric ring. Thus there is formed an effective apparatus, antagonistic to the pyloric sphincter by means of which the pyloric orifice may be dilated when the nervous mechanism concerned in inhibition is stimulated. This enervation is through the splanchnic nerves. There is then both a constrictor and a dilator of the pylorus.

The circular muscle fibers of the stomach are not continuous directly with those of the duodenum. On the contrary, at the pyloric orifice they become very much increased in number and form a thick ring or sphincter which is separated from the circular muscular coat of the duodenum by a fibrous septum.

The pyloric orifice is directed horizontally backwards, and to the right, when the stomach is empty, but when the stomach is full it looks almost directly backwards or even slightly to the left side. The body of the pylorus rests on the neck of the pancreas below and posteriorly and is overlapped by the liver above, and anteriorly. The pylorus is lined with mucous membrane continuous with that of the stomach and contains short, simple, branched, tubular glands, which are not as tortuous as those of the stomach. The transition from fundus to pylorus is not abrupt, but is marked by a transitional border zone in which fundus and

pyloric glands are intermingled. The stroma in which the glands are imbedded is greater in amount in the pylorus and the glands are more widely separated. There is considerable infiltration of the stroma with lymphoid cells. The muscularis mucosa generally consists of an inner circular layer and an outer longitudinal layer of smooth muscle fibers. The submucosa consists of loosely arranged connective tissue containing blood vessels and the sympathetic ganglion cells forming the plexus of Meissner. The serous coat consists of loosely arranged connective tissue.

The blood supply of the pylorus is derived from the left gastric branch of the celiac artery and from the hepatic branch of the celiac through the pancreatico-duodenal.

The nerve supply is derived from the plexuses of sympathetic nerves around the superior mesenteric artery. From this source they run to the myenteric plexus, or plexus of Auerbach, situated between the circular and longitudinal muscular fibers, from which the nervous branches are distributed to the muscular coats. From this a secondary plexus, the plexus of the submucosa, or Meissner's plexus, is derived, and is formed by branches which have perforated the circular muscle fibers. This plexus supplies the muscularis mucosa and the mucous membrane. The nerve bundles of the submucous plexus are finer than those of the myenteric plexus. The vagus nerve is motor to the pyloric sphincter while



the sympathetic supplies the longitudinal muscle layer of the pylorus whose contraction aids in dilating the canal. The sympathetic is also inhibitory. The relationship between the enervation and movements of the pylorus has not been definitely worked out.

#### PHYSIOLOGY

The original theory of Cannon and Pavlov, (17) claimed that the pyloric sphincter was controlled mainly by the acidity of the gastric contents. According to their theory the acid chyme in the stomach stimulated the opening of the pyloric valve and its presence in the duodenum, after passage through the pylorus, stimulated its closure.

This theory has been abandoned at the present time. It has been definitely proven that the pylorus is not controlled by the gastric acidity. Campbell (18) has shown that there may be a complete achlorhyeria with a normal emptying time of the stomach. The placing of acid in the duodenum had no effect on the pyloric sphincter.

From the study of serial cardiographs with graphic records, produced by means of a balloon inserted into the stomach, Carlson (19) found that the passage of chyme into the duodenum coincided with the peristaltic waves traveling over the stomach. By further experimentation on a dog, it was found that ingested water

passing into the duodenum was neutral in reaction. These experiments disprove the "acid control theory" and show that the pyloric sphincter opens in response to the peristaltic waves in the stomach.

The exact relationship between the nervous system and the movements of the stomach and pylorus has not been thoroughly worked out. In a stomach which has been removed and kept in warm saline solution, definite rhythmic contractions of the fundus and pylorus have been observed. The impulses must therefore, originate in a local nervous system, probably the plexus of Auerbach situated between the layers of the muscular coat. Auerbach's plexus has been severed by Cannon (18) and the peristaltic waves still pass through. The origin of these impulses may be considered as myogenic<sup>AS</sup> in the case of heart muscle.

The vagus nerve is considered as motor to the pylorus. Stimulation of the vagus causes first an inhibition and then a vigorous contraction of the sphincter, and has the same action on gastric peristalsis. The sympathetics, when stimulated, produce an inhibitory action on peristalsis and tend to relax the pylorus. The exact relationship between the nervous system and the movements of the stomach and pylorus is not definitely known.

## PATHOLOGY

The pathologic picture of the pylorus described by numerous authors has been quite uniform and there have been relatively few discordant opinions. The essential lesion is one of hypertrophy and hyperplasia of the musculature of the pylorus especially of the circular muscle. Other changes are nearly all secondary to the obstruction produced by this muscular anomaly and the added spasm.

The pyloric canal has the appearance and consistency of a solid cylinder about two to five centimeters in length. The tumor varies in size from that of the distal phalanx of the little finger to that of the thumb, and the diameter is generally greater in the middle than at the extremities giving the tumor a rather fusiform or olive shape. The tumor has a covering of normal glistening peritoneum and is sharply demarcated from the distended stomach above and the empty duodenum below. It is freely movable with no peritoneal adhesions. On longitudinal section it presents a dense, firm, uniform structure, down to the mucosa. Microscopic examination reveals a definite hyperplasia of the circular muscle layer increasing the thickness by two or three times. Normally in children under three months the thickness is not greater than 2.5 mm. the average being 1.6 mm. In the hypertrophic pylorus at this age the circular muscle layer shows

a thickness of from three to seven mm. There is often edema of the submucosa while the other layers remain unchanged. The sphincteric ring is slightly, if at all, involved in the hypertrophy the hypertrophy being limited to the circular muscle layer of the pyloric canal. Inflammatory changes in the pylorus are rarely seen, and when present are probably secondary to the retention of food, and traumatism resulting from hyperperistalsis.

The walls of the stomach are generally somewhat hypertrophied and the cavity dilated. Wollstein (20) reports a series of twenty five necropsis on infants with hypertrophic pyloric stenosis. She found in all, that the stomachs were dilated, often twice the normal size.

The mucous membrane generally showed evidence of some degree of gastritis and there was slight hypertrophy of the musculature. These changes in the stomach are secondary resulting from distention, by accumulated contents and hyperperistalsis.

#### SYMPTOMS

The symptoms of the condition present a well defined and uniform clinical picture. The first cases of congenital hypertrophic stenosis of the pylorus described by Blair in 1717 and those of Armstrong and Beardsley which have already been mentioned, present a symptomatology almost identical with the cases of modern observers.

Bolling (21) in a study of four hundred fifty four cases, states that the condition usually occurs in breast fed male infants, frequently the firstborn, and that the ratio between male and females is four to one. The frequency in male infants is stated as one in every two hundred. In this series of cases the symptoms began in the second to the fourth weeks. Onset after the fifth week was rare and after the eighth week exceptional.

The onset is usually abrupt, a previously healthy baby will suddenly begin to vomit. Steen(22) in a recent article, states that "in a wasted child there are two symptoms which, associated together, are of outstanding value in suggesting pyloric stenosis. They are, vomiting, and constipation. The combination of vomiting with constipation is as rare in infancy as that of vomiting and diarrhoea is common".

The vomiting is characteristic, being forcible and projectile, sometimes the food is shot out of the mouth to a distance of several feet. The baby accepts food eagerly and is apparently hungry. After each nursing it may vomit and the amount is usually large, frequently the entire contents of the stomach. If the vomitings are infrequent, the amount ejected at one time is often larger than the amount of the last feeding. There is no apparent nausea associated with the vomiting.

The vomitus is bile free and usually shows hyperacidity. It is often blood streaked. The blood is due to simple mechanical violence resulting from distention and hyperperistalsis of the stomach.

Shortly after the onset of vomiting constipation occurs, which in advanced cases reaches a degree that may properly be described as absolute. The meager stools that can be obtained by enemas or suppositories are typical starvation stools, and consist mainly of bile stained with mucus. The persistent absence of bile rules out the possibility of obstruction below the duodenum. There are frequently small amounts of blood present in the stools.

As a result of the pyloric obstruction and vomiting, an absorption of all food and liquids is almost completely stopped. This leads to a marked loss in weight, diminution in the output of urine, and dehydration.

The loss in weight usually occurs after a preliminary normal gain, depending, of course, of the time of onset of the primary symptoms. This loss is a constant symptom and its rapidity is the best guide to the seriousness of the case. In a series of cases (I) the loss varied from fifteen to twenty percent of the total body weight in ten days, to a total loss of as high as thirty five percent, the loss varying directly with the

degree of obstruction.

As the result of fluid loss, the urine early becomes concentrated and decreased in amount, and may deposit a brick red uric acid sediment.

The child is usually more or less fretful, other than this, there is nothing characteristic in the general picture. The fretfulness is probably due to hunger more than anything else. The temperature is always normal unless there is some secondary infection, or the condition is far advanced. In the later, fever is present if the dehydration has reached such a degree as to cause an incontinant fever. Henske (23) reports a case of this character in which the fever was promptly reduced by the administration of intra-peritoneal normal saline solution. Thus demonstrating that the temperature was due to loss of fluids.

In 1929, Graham and Morris (24) in a study of fifty cases of hypertrophic stenosis of the pylorus, demonstrated definite changes in the chemistry of the blood. These changes consisted in relative degree of ~~alkalosis~~ with a high carbon dioxide combining power of the blood plasma. This they explained as being due to a loss of chlorides by vomiting of the gastric juice. This in turn leads to an increase in the carbon dioxide combining power of the blood plasma to make good the deficiency of acid radicles. As the result, the respiratory rate is reduced

and with this an associated general lethargy occurs. With intravenous injections of sodium chloride solution, the experimenters were able to bring the carbon dioxide content to normal, and increase the respiratory rate.

From these clinical and laboratory findings it was concluded that pyloric stenosis in infancy is associated with an alkalosis presumably produced by a loss of the chlorine acid radicle. The loss of chlorine is definitely related to the severity of the vomiting, and the carbon dioxide content varies inversely with the blood chlorides.

The symptoms may then be briefly summarized. A sudden onset of projectile vomiting, followed by constipation and diminution in the output of urine and an early loss of weight, are the classical symptoms of hypertrophic pyloric stenosis. As already discussed, there may be an alkalosis present resulting in depressed respiration and lethargy. The alkalosis is peculiar in that it is due to a loss of chlorine acid radicles, causing a compensatory carbon dioxide combining power of the blood plasma. Symptoms of inanition are dependent upon the degree of dehydration.



## PHYSICAL FINDINGS

Early in the course before there is much loss of weight the child appears perfectly healthy. After the condition has persisted for some time there is more or less emaciation and dehydration present. The greater the obstruction the greater the degree of emaciation and dehydration.

The abdomen presents a peculiar conformation (I). The upper part is full, tense and bulging and is made more prominent by a marked retraction of the hypogastric region, presenting a somewhat funnel shaped appearance. Slowly moving peristaltic waves cross the upper abdomen from left to right, their size and frequency being increased by the ingestion of food or water. Similar waves occur in congenital atresia or stenosis of the duodenum, but the presence of symptoms from birth and the bilious character of the vomitus usually serve to distinguish these anomalies. Slight atypical waves may be seen in apparently normal infants. There is no muscle rigidity present as in cases of an inflammatory condition. Together with this lack of rigidity, and the thinness of the abdominal wall resulting from loss of weight, palpation of the viscera is quite easy.

In nearly every case reported by Downes Richter and Bolling,

it has been possible to palpate a definite tumor at the pylorus. This is regarded as one of the important diagnostic signs. It can be felt lying between the right costal arch and the umbilicus and as a freely movable solid tumor with a definite outline. It is usually olive shaped. The tumor is most easily palpated after the stomach has been emptied by vomiting or by means of a soft rubber catheter. The child is held comfortably with the trunk slightly ventrally flexed and the thighs flexed on the abdomen. This relieves muscle tension. A sugar pacifier may be given to keep the child quiet.

The pylorus may usually be made out to the right of and just above the umbilicus although some variations from this rule are common. The tumor is an oblong, movable mass about the size of the distal phalanx of the little finger and imparts a sense of almost cartilagenous hardness through the abdominal wall. The consistency varies with the contraction and relaxation of the muscle composing it. In the four hundred fifty four cases reported by Bolling, the tumor was palpated in every instance but one. During the same period there were three cases in which the ante-operative diagnosis of a tumor at the pylorus proved incorrect. From this one can readily see the value of tumor palpation as a diagnostic sign.

#### XRay Findings

X Ray examination made after a barium meal reveals a distended stomach with very little or no passage through the pylorus. The majority of observers believe that this is mainly a

confirmatory and that a diagnosis may be made without the aid of roentgenography.

### DIAGNOSIS

If the condition is kept in mind the diagnosis may be made promptly in almost every instance, and is not a matter of opinion but is demonstrable. A careful history, if obtained from an intelligent mother or nurse is valuable and often typical. Projectile vomiting occurring in a normal infant, followed by marked constipation and loss of weight, is characteristic. A palpable pyloric tumor and gastric peristaltic waves in combination occur in no other condition. The roentgenographic examination confirms the diagnosis but is not essential in making it.

#### Differential Diagnosis.

In diagnosing the condition there are other possibilities which must be ruled out. These are (a) Simple stenosis of the pylorus. (b) Congenital intestinal atresia (c) Cardio-spasm and esophageal atresia, and (d) Congenital retroperitoneal hernias.

(a) Simple stenosis of the pylorus is usually present from birth and is due to a spasm of the pylorus without hypertrophy. The symptoms are *much* the same as in congenital hypertrophic stenosis, but to a much less degree. The loss of weight is not as

marked and the stools are not of the starvation type. There is no palpable tumor felt at the pylorus although slight peristaltic waves can be seen. There are remissions of vomiting which are not found in true hypertrophic stenosis. The patient responds to atropine.

(b) It is differentiated from all congenital intestinal atresias on the basis of an interval after birth before symptoms develop. In cases of atresia the symptoms are present from birth. The time factor also serves to eliminate atresia of the esophagus. The vomiting in esophageal atresia is very mild.

(c) Cardio spasm may be present from birth or may develop later. It is always associated with vomiting either during active swallowing or immediately following it.

(d) Congenital retroperitoneal hernias have presented obstructive symptoms, in some instances from the time of birth, in others beginning some time after birth. Retroperitoneal hernias with obstruction produce symptoms quite similar to hypertrophic stenosis of the pylorus. A mass may be palpated in the upper right abdominal quadrant, but it is larger and softer than a pyloric tumor. The vomitus as a rule contains bile which is not present in hypertrophic stenosis. There is usually blood in the stools which is characteristic of intestinal obstruction.

Any of these possibilities may be ruled out by roentgenological examination.

#### TREATMENT

The treatment of the condition may be grouped under two main headings namely surgical and medical. The tendency at the present time, is to favor the surgical treatment. The mortality has been greatly reduced and the results permanent.

##### Surgical

It is interesting to trace the evolution of the operative treatment through the more complicated methods such as jejunostomy, pylorotomy, division of the pyloric canal, gastro-enterostomy, and pyloroplasty down to the simple extra mucous partial pyloroplasty. This procedure is now known as the Fredet Rammstedt operation. The first successful operation a gastro-enterostomy was performed by Lohrer (6) in 1898. This procedure became the operation of choice for the next fifteen years, but the mortality was extremely high. In 1907 Fredet (6) performed the first successful pyloroplasty by means of an incision through the pyloric wall down to the mucous membrane. Weber (6) in 1910 emphasized the same procedure. In 1912 Rammstedt (6) called attention to the advantages of this operation but advised the omission of the transverse suture and suggested leaving the wound gapping. This simple procedure now known as the Fredet Rammstedt operation has revolutionized the treatment of the condition.

The operation should not be carried out as an emergency measure. Infants in reasonably good condition, who are not dehydrated, may be operated upon within twenty four hours of admission. Even in such patients a hypodermoclysis of three percent glucose in salt solution is usually given about twelve hours before operation.

Where symptoms have existed for a long time and the infant is emaciated and dehydrated, operation should be delayed for two or three days. The body heat should be maintained and fluids and nourishment replaced by intravenous injections of glucose, normal saline solutions and transfusions.

The anaesthetic of choice is ether because of the relaxation obtained. In infants whose nutrition and general condition is poor, local anesthesia is preferred, the line of incision being infiltrated with a one percent procain solution.

The Predet Rammstedt operation consists in making an incision along the convex border of the pylorus parallel with the lumen. The incision extends from the duodenal end well on to the wall of the stomach and varies in length from 1.5 to 2.5 centimeters. The incision involves the superficial muscle fibers of the pyloric canal, a pair of forceps is then used to gently spread the remaining

undivided fibers down to and exposing the mucous membrane. The mucous membrane should protrude freely into the incision. The abdominal wall is then closed, no attempt being made to cover the wound with omentum.

Post operative treatment consists largely in nursing care. The child should be kept warm and if a general anesthetic has been used, the head should be lowered to prevent aspiration pneumonia. Richter (I) advises the subcutaneous injection of normal salt solution for twenty four hours following the operation. Water by mouth may be started in a few hours the amount being gradually increased, and breast milk if available may be given within twelve hours. It is particularly desirable that the mother's milk should be used and retained in the after-care of the child.

The results of surgical treatment depend largely upon early operation. From January 1920 until 1925 Bolling (21) has operated upon two hundred seventy nine patients, consisting of four hundred private patients and two hundred thirty nine ward patients. The private patients represent early diagnosis and operation resulted in a mortality of only 2.5 percent. The ward patients represented cases, the majority of which were quite far advanced, the mortality in this instance was over fifteen percent.

This is quite a striking contrast, and shows the urgency of early diagnosis and treatment. Of a total number of five hundred eighty cases operated upon by Bolling and Downes, employing the Fredet-Rammstedt technique, the mortality has been less than four percent. This data shows the merits of the Fredet-Rammstedt operation.

#### Medical

Before surgical treatment was introduced, numerous non-operative methods were employed for the relief of this condition. There are still men who advocate surgery, only after medical treatment has been given a fair trial.

Gastric lavage was used almost universally in the beginning of medical treatment. This consisted of washing the stomach with a one percent sodium bicarbonate solution, repeated twice daily about two hours after a feeding. The purpose of which was to relieve irritation of the gastric mucosa and reduce peristalsis. With this procedure the mortality was high and is very seldom used at the present time.

In 1918, Sauer (25) recommended the use of thick Farina feedings in the treatment of the condition. In a series of twelve cases with typical symptoms and physical findings of a tumor at the pylorus, eleven recovered under this treatment,



the remaining one dying of bronchopneumonia.

The purpose of thick feedings is mainly to produce mechanical dilatation of the pylorus. Thick food is less easily regurgitated, and with the vigorous peristaltic contractions, produces a mechanical dilator. Sauer recommends a thorough trial of this treatment before surgical means are resorted to.

In 1919 Haas (10) advocated the use of atropine in the treatment of pylorospasm and congenital hypertrophic stenosis. His theory which has been discussed earlier in this paper under "etiology", states that the condition is due to a general state of hypertonicity. The pyloric hypertrophy resulting from an overactivity of the vagus nerve, known as vagotonia. This led to the use of atropine, in the belief that if properly used it will effectively cure the condition.

The atropine used consists of the freshly prepared drug in the form of atropine sulphate and made into a one to one thousand solution. He advocates enormous doses reaching from one fiftieth to one twenty fifth grain in twenty four hours. The first dose is one drop of the one to one thousand solution, to be put into the feeding. If there is no idiosyncrasy to the drug, the dose at the next feeding is two drops. This increase in dosage is con-

tinued until the child is getting from one fiftieth to one twenty fifth of a grain in twenty four hours. If toxic symptoms occur the drug is withheld for a few feedings and then continued.

With this use of atropine the symptoms have entirely disappeared in from a few weeks to several months. Haas therefore advocates the use of atropine in every case, continuing its use indefinitely so long as symptoms are present. This is demonstrated when the dosage is lowered or when it is withheld entirely.

In a majority of the cases treated by Haas the symptoms were typical of pyloric obstruction and a well defined tumor could be palpated in the region of the pylorus.

Sauer (26) in 1929 introduced the use of luminol in the treatment of pyloric spasm and pyloric stenosis with apparently good results. Simple cases of pyloric spasm responded better than true hypertrophic stenosis. Barbour (27) in a further study of the luminol treatment, favors it over the use of atropine. The tolerance to luminol does not vary as greatly as does the tolerance to atropine. In two cases of hypertrophic stenosis, luminol was given before and after the Rammstedt operation. Each infant was enabled by its use to retain enough food to permit sufficient improvement in its nutrition for the surgical risk.

As yet, the use of luminol has been too limited to arrive at any definite conclusions.

In 1928, Wiener (28) found that good results could be obtained using a combination of medical, dietary and X Ray treatments. He believes in a restriction of operative indication. Six cases are reported in which X Ray treatment was very satisfactory. One or two treatments were sufficient to relieve the distressing symptoms and there was apparently no return of the vomiting.

If this procedure can be proven of definite value, it will be a very welcome addition in treatment of pyloric obstruction. Any method which proves an equal substitute for surgery is the method of choice.

#### COMPARISON OF MEDICAL AND SURGICAL TREATMENT

It is quite difficult to say that one case should be treated medically and the other surgically. If the patient does not respond promptly to medical procedures, delay in operation may prove fatal. Goldbloom and Spence (29) have shown that when the symptoms have lasted less than four weeks, the mortality is only one third as great as when they have lasted four weeks or more. The mortality increases in direct proportion to the amount of weight lost.

The medical treatment as a rule, has to be maintained over a long period of time and the child sometimes becomes so emaciated and dehydrated that it is no longer a surgical risk.

This, however, is not true in all cases. Some respond promptly to medical treatment and permanent relief of all symptoms results. Therefore, if after a fair trial, medical methods bring about no improvement, surgery should be resorted to.

In a study of twenty-One infants, twelve representing true cases of hypertrophic stenosis and seven representing cases of pylorospasm, Greer (30) found that medical treatment was successful only where pylorospasm was present. Greer states, "I have never obtained a cure of pyloric vomiting by medical and dietary means in any case in which the diagnosis of congenital hypertrophic pyloric stenosis was not open to serious question".

In a discussion of Doctor Greer's paper Doctor Helmholtz of Rochester, Minn. claims that he has definitely cured cases of pyloric stenosis by atropine and thick gruel feedings. In infants dying after a permanent cure by medical treatment, there was still a demonstrable tumor present.

These articles show the controversy existing as to the comparative value of medical and surgical methods of treatment.

## CONCLUSIONS

1. The first case was described in 1717 by Patrick Blair.
2. The exact etiology of the condition is yet unknown.
3. The pathology consists in a hypertrophy and hyperplasia of the pyloric circular muscle fibers. The hypertrophy is probably congenital and the obstruction results from an added spasm occurring in early infancy.
4. There appears to be a direct relationship between pyloric stenosis and Vitamin B. deficiency.
5. In a number of cases, there has been an associated allergic condition. Dietary elimination of the specific allergin and medical treatment gave relief from the symptoms.
6. The physiology of the pylorus is not definitely known. The present theory being that the pylorus opens in response to gastric peristalsis.
7. The symptoms are typical and early diagnosis is not difficult.
8. Early treatment is essential.
9. Medical treatment is still of questionable value in this condition.
10. The Fredet Rammstedt surgical procedure is the treatment of choice.

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