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The evolution of the treatment of pyloric stenosis



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AUTHORS

Chinwendu Onwubiko, MD, PhD

Barry R. Berch, MD, FACS

David E Sawaya, MD, FACS

Don K. Nakayama, MD, FACS

Christopher J. Blewett, MD, FACS

Department of Pediatric Surgery, University of Mississippi Medical Center, Jackson, MS

CORRESPONDING AUTHOR

Chinwendu Onwubiko, MD, PhD

Department of Surgery

University of Mississippi Medical Center

2500 North State Street

Jackson, MS 39216

601-984-5101

conwubiko@umc.edu

Once associated with a high mortality when first described in the early 18th century, surgery for infantile hypertrophic pyloric stenosis today is commonplace and all infants are expected to make an uneventful recovery. How surgery evolved over time to the present standard operation, pyloromyotomy, is a story of surgeons familiar with adult conditions gradually discovering a condition specific to infancy and arriving at a correspondingly unique operation.

Patrick Blair, a Scottish surgeon, described a case of an infant who appeared to have pyloric stenosis in 1717.¹ Harald Hirschprung made the first specific description of pyloric stenosis in 1888: "the mucosa showed six ledge-like parallel columnae protruding along the entire length of the canal. These ledges form a rosette, which projected into the cavity."² Pyloric muscle hypertrophy was identified as the cause of gastric obstruction. Of note, Hirschprung's first two cases were in female infants, though pyloric stenosis is 4-5 times more likely in males.^{3, 4}

Before Frédet

Early authors believed that the muscle hypertrophy was due to pyloric spasm. Management therefore proposed methods to decrease gastric contractions. Approaches included application of electrical current and antispasmodic medications such as atropine and belladonna. Other interventions were neutralization of gastric acid with bicarbonate solution lavage, and refeeding of emesis to ameliorate dehydration and electrolyte imbalances. These measures were frustrating to apply, but were the only means to nurse infants over the course of the six months or so required for gradual resolution of the condition, its natural history. Many infants died before then from dehydration and inanition.^{2, 5}

Surgery became an option once it became accepted that pyloric hypertrophy, not spasm, was the cause of the pathology. (One century late molecular technology found the pyloric musculature in infants with pyloric muscle lacked nitric oxide synthase, the enzyme involved in the release of nitric oxide, a mediator of smooth muscle relaxation.⁶) The infants were poor candidates for surgery, inflicted with dehydration, malnourishment, and severe acid-base and electrolyte imbalances. Open drop anesthesia made depth of anesthesia uncertain, made worse by induction in a baby with a full stomach and gastric obstruction. The distended stomach was especially troublesome since once opened, discharged its contents into the field and peritoneal cavity. The contamination was a potential disaster in the era before antibiotics and routine preoperative drainage through a catheter passed from the nose or mouth. Mortality for surgery was correspondingly high, up to 61 percent in early reports.⁵

Lobker first employed gastroenterostomy successfully in 1898 in infants^{3, 5}, an operation first employed by Billroth's assistant Woelfler for obstructing cancers of the stomach in adults in 1881⁵. The following year James Nicholl dilated the pylorus from an incision in the stomach, which he paired with a gastroenterostomy.^{3, 5} He adopted the procedure from Loreta from Italy, who used the operation in 1884 for the treatment of adults with pyloric strictures.⁷

Addressing gastric outlet obstruction from acid-peptic disease in adults Heineke in 1886 and Mikulicz-Radecki in 1887 independently devised their classic operation, the longitudinal incision across the pylorus and its closure in a transverse direction. On June 10, 1902, Clinton Dent in London applied the operation to infants with pyloric stenosis. In his report he noted that some of his contemporaries recognized the difficulty of closing a longitudinal incision in the required transverse direction, given the thickness of the pyloric muscle. Dent claimed that he did not encounter any difficulty in performing a Heineke-Mikulicz procedure on infantile disease. In fact, he claimed, the procedure was easier in infants than in the pyloric

inflammation found in adults. Among the 21 patients in his series there were some who underwent gastroenterostomy and resection of the pylorus. He had some successes, but his overall mortality was near 50 percent.⁸

Frédet

Pierre Frédéric is widely credited as performing the first “extramucosal pyloroplasty” on October 12, 1907 in which the mucosa was left intact during division of the pyloric muscle.² One month earlier he had tried a Heineke-Mikulicz pyloroplasty on a full-thickness incision of both muscularis and mucosa. His sutures cut through the duodenal mucosa, forcing him to attempt to suture the layer to the antral side of the incision. After the operation the infant began to vomit blood, and died the next day. The lesson learned, Frédéric resolved to stay out of the lumen in his next operation, completing the pyloroplasty without violating the submucosal layer. Five years before Dent had brushed off a suggestion that such an operation might be successful.⁸ Frédéric disagreed; just a month later before the Societe Medicale des Hopitaux de Paris he reported:

One considers a pyloroplasty, an operation which seems a priority, the easiest and least dangerous with an incision about 2 cm long on the axis of the pylorus in the middle of the superior aspect. This longitudinal incision carries through the peritoneum and the muscularis to the exclusion of the mucosa. The bistoury cuts a white tissue, edematous and very hard, creaking under the instrument, having every appearance of certain uterine myomas. The incision cuts entirely through the sphincter to a depth of several millimeters, and the lips of the wound are gently spread. A series of sutures of linen, placed according to the method of Heinecke and Mikulicz, transform a longitudinal wound into a transverse wound, a plastic procedure, which manifestly enlarges the pylorus. The sutures, to the number of 6 or 7, take the entire thickness of the muscle mass and are tied successively to avoid their cutting through.²

In part due to avoiding free spillage of gastric contents, mortality fell to 17%.^{2,3,7} The next year in 1908 Weber from Germany performed an identical operation.⁷

James Nicholl, who previously had tried both Lobker's operation and gastroenterostomy⁷, developed an extramucosal pyloroplasty before Frédéric, the former's first case taking place almost three years before the latter's first success. Nicholl published a series of six a year before Frédéric's presentation to his Parisian colleagues. He used a V-Y pyloroplasty oriented transversely. At first he dilated the pylorus through a separate gastrotomy, but eventually found that he could open the pyloric channel by grasping opposite sides of the incision and pulling them apart.⁹ Taylor noted that Nicholl was the forgotten figure in the history of pyloric stenosis. “Nicholl of Glasgow,” he wrote, “seems never to have obtained rightful credit for his work.”⁷

Frédéric doubted the reliability of his operation. He felt that it should not be used in all situations, especially in those patients with large pyloric muscle where he could not close the muscularis. He did not trust leaving the mucosa exposed, so he would instead revert to a gastroenterostomy in those situations. In a 1921 review 9 of his cases had received gastroenterostomies.²

Frédéric spent a great deal of his career studying pyloric stenosis. Among his lasting contributions was the phrase “projectile vomiting” as a trigger phrase associated with pyloric stenosis. He also described the nonbilious emesis and visible peristalsis as signs indicating the diagnosis. From his hard experience he learned the necessity of preoperative gastric decompression, keeping the baby warm during and after operation, and maintaining hydration through clysis (subcutaneous injection of fluid).²

Dent was probably the only surgeon willing to publicly state that closing a pyloroplasty was easy. All other surgeons tackling pyloric stenosis, especially Frédéric and Nicholl, had problems getting the thick muscularis to close in a transverse direction without sutures cutting through the tissue.

Ramstedt

On August 23, 1911, Dr. Conrad Ramstedt began an operation for pyloric stenosis with the intention of performing a pyloroplasty. He was already nervous before he started; it was his first such case, and the patient was the son of a famous nobleman.⁷ After he weighed his options he decided on pyloroplasty as recently described by Frédéric, despite not having seen nor done one.¹⁰ Ramstedt recalled the operation more decades later:

After I had split the tumor down to the mucosa for a distance of about two cm, I had the impression that the stenosis had been relieved. I still tried, however, to accomplish the previously described plastic procedure by transverse suture of the muscle edges. However, the tension on the sutures was so very strong that the first one cut through immediately. Then the thought shot through my head, 'A plastic alteration of the cut edges is completely unnecessary; the stenosis seems to be already relieved by a simple splitting of the pyloric muscle and coincidentally the spasm as well, which is the characteristic basis of the disease.' I did not complete the plastic operation on the muscle which had been originally planned but left the cut gaping, covering it with a tab of omentum for safety's sake and ended the operation. The little one vomited a few times for the first few days which I attributed to the sutures placed at the beginning, but he recovered promptly and completely to the great joy of his parents .

His next case came the next year on June 18, 1912. Using the same operation he successfully treated the infant son of parents who had already suffered through pyloric stenosis in

three other children, two of whom had died.⁷ After Ramstedt reported his cases at the Natural Science Assembly at Münster in September, his operation became widely adopted. In the U.S. series of respectable size began to appear by 1916. It was not until the end of the war approached in 1918 that the operation was attempted in England. The initial British attempt was successful and surprisingly easy to perform, being completed in just seven minutes.⁷

Conclusion

The history of treatment of pyloric stenosis is doubly gratifying, the story of an operation elegant in conception and so effective that it has been called, "the most consistently successful operation ever described."⁴ Surgeons tried to apply operations designed for adult pathology, only to arrive at one that was unique to a condition seen only in infancy. For a condition that was once fatal in nearly all cases, recovery from pyloric stenosis is today the norm, after only a brief operation and a short hospital stay. Now commonplace, pyloric stenosis is one of the overlooked triumphs of pediatric surgery.

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