CONGENITAL HYPERTROPHIC PYLORIC STENOSIS*

EDWARD J. DONOVAN, M.D.

NEW YORK, N. Y.

IN FORMER PAPERS on congenital hypertrophic pyloric stenosis published in 1932 and 1937, I reported a total of 243 consecutive cases operated upon by me at the Babies Hospital in New York with only one death. The purpose of this paper is to bring up to date my personal series of cases at the above hospital, a series which, with the 245 cases upon which I have operated with two deaths since 1937, now comprises 507 cases. We have made no important changes in the treatment, always using the Fredet-Rammstedt operation and emphasizing the immediate preoperative preparation and postoperative care. The success of modern surgical treatment, in which a cure can be permanently and easily achieved by a simple operation continues to prove its superiority over extended medical treatment with its prolonged and uncertain results. With a mortality of 1.8 surgery can be more vigorously recommended than when the mortality was 35 per cent. Parents can now be assured not only that their baby may be operated upon successfully and that he will be retaining all feedings and gaining weight 10 days after operation, but also that he will have no stomach trouble later in life as a result of his pyloric stenosis in infancy.

Etiology.—In spite of the many theories which have been achieved, the etiology is no clearer today than it was in 1887 when Hirschsprung presented two of the earliest cases on record. There are still many phases of the disease and its cure which are unexplained, the chief of these being whether the hypertrophy of the circular muscle precedes or follows the pylorospasm. Others are the occasional spontaneous cure, the disappearance of the tumor about seven weeks after the Fredet-Rammstedt operation, its persistence throughout life in patients who have had posterior gastro-enterostomies, the hypertrophy of the circular muscle coat, the occurrence of the condition about seven times more often in boys than in girls, and its frequency in the first child of a family.

It is our belief that the tumor is congenital in origin, that the pylorospasm follows the hypertrophy and that pylorospasm is responsible for the onset of symptoms between the second and fifth weeks of life. This belief is supported by finding well-marked tumors in each of two premature infants and by the absence of correlation between the size of the tumor and the age at which operation is performed. Fully developed tumors have been found in many babies upon whom we have operated a few days after the onset of symptoms. The variation in the severity of the symptoms seems to be due rather to the amount of pylorospasm present than to the size of

* Read before the American Surgical Association, April 2–4, 1946, Hot Springs, Virginia.

the tumor, since the largest tumor is often present when the onset of the symptoms is least severe.

Pyloric stenosis has occurred in one of twins, the other twin being perfectly normal, and in both babies when the twins were identical. We have found it in all races and nationalities with a slight predominance in the children of Jewish parents, and we have had about two colored babies in each hundred cases. We have not been able to demonstrate a seasonal incidence.

Pathologic Anatomy. — Operative treatment of pyloric stenosis has made it possible to confirm the pathologic findings as described by the earliest writers. The most striking feature is the firm, almost cartilaginous, tumor found at the pylorus. This tumor, which completely encircles the pylorus, is about 3 cm. long and 1.5 cm. in diameter. It is freely movable and projects into the duodenum as the cervix does into the vagina, gradually blending into the pyloric antrum at the gastric end. The term "scirrhous" as applied to this tumor by the earliest writers is quite appropriate. When stimulated, the tumor becomes blanched and firmer in consistency, and when the circular muscle is cut, it is found to be 5–10 mm. in thickness. The stomach is normal except for compensatory hypertrophy and dilatation. Cross-section reveals the lumen of the pylorus almost completely closed and the hypertrophy confined to the circular muscle coat. Microscopic study confirms the site of the hypertropy, and shows all other structures to be normal. The pathologic anatomy in cases of long-standing does not differ from that in cases of short duration except that the stomach is hypertrophied and dilated to a greater degree.

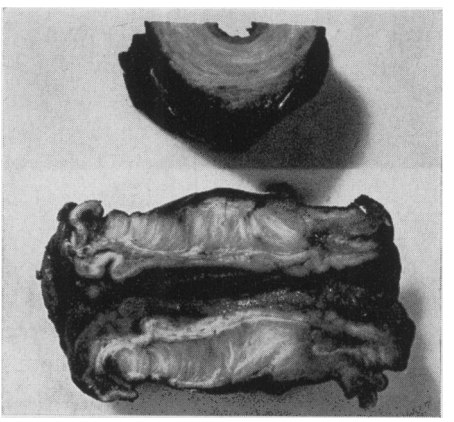

FIG. 1.—Pylorus removed in a patient 29 years of age. Gastro-enterostomy performed in infancy for pyloric stenosis. Pylorus looks much the same as these tumors do in infancy. Microscopic section shows only hypertrophy of the circular muscle coat; width 5 cm.

Clinical Course.—The clinical picture of pyloric stenosis is impressively uniform. Vomiting during or after each feeding is always the first symptom. It may start abruptly, but it usually begins as a regurgitation. The quantity vomited may be small at first, but it increases later, and it may amount to considerably more than the previous feeding due to gastric secretion and to the ever-present gastric retention. The vomiting soon becomes projectile and forceful enough to eject the stomach contents several feet, but it cannot be painful or accompanied by nausea since a complete feeding will often be taken immediately thereafter. The vomitus is never bile-stained, but it may contain mucus and bright blood or coffee-ground material resulting either

from the associated gastritis or from the rupture of a mucosal vessel during the act of vomiting. The baby may lose enough fluid by vomiting to become extremely dehydrated, to lose weight rapidly, to be constipated and to pass only small amounts of concentrated urine.

The course of pyloric stenosis is liable to be rapidly progressive; it is not unusual for these babies to lose from 30 to 50 Gm. in weight daily if the condition is not recognized and treatment instituted. If fluid is not restored, one may expect death from dehydration and starvation in about four weeks' time. Occasionally one sees a baby in whom the progress of the disease is much slower even though the tumor is well defined, and it is in this type of case that medical treatment may be successful.

Diagnosis.—The diagnosis depends entirely upon a history of the course as described and upon palpation of the pyloric tumor which is present in every true case, and which is pathognomonic, being found in no other condition. Gastric peristaltic waves, although always present, their intensity depending upon the duration of the obstruction, cannot be considered diagnostic since they occur in other conditions. The waves start at the left costal margin and pass across the epigastrium toward the right side. It is not unusual to see a second wave start before the first has completely disappeared. In a long-standing case, in which there has been a considerable loss of weight, the waves are easily seen, and the outline of the full stomach is sometimes evident through the thin abdominal wall. While waves are most obvious after the baby has had a feeding or when the stomach is full, the tumor is felt most easily when the stomach is empty and the baby is relaxed with a sugar pacifier. As we believe that the tumor can be felt in every case, we do not operate until it has been felt. If some such rule is not established, many patients will be operated upon who do not have pyloric tumors. The tumor, which feels like a small olive, is found most frequently in the right upper quadrant to the right of the lateral border of the rectus muscle and about 4 cm. above the umbilicus. If the tumor lies beneath the liver or the right rectus muscle, it is difficult to feel, and a bulge of rectus muscle between the linea transversa may be mistaken for it, particularly if the abdomen is not well relaxed. If one presses the fundus of the stomach toward the midline with the left hand while palpating for the tumor with the right hand, many tumors will be brought out where they can be felt easily. The tumor may often be found at the point over the right epigastrium where the peristaltic wave disappears. It is in many instances surprisingly superficial. Although diagnosis may be made roentgenologically, we believe this to be not only unnecessary but actually contraindicated since it greatly increases the baby's postoperative discomfort if all the medium is not removed.

Preoperative Treatment.—Restoration of the fluid lost by vomiting before operation is, without doubt, the most important factor in the present low surgical mortality. This operation should never be considered an emergency, and no baby should be operated upon until he is completely hydrated. It is perfectly safe and often necessary to spend several days

in preparing a baby for operation. Because many of these babies have alkalosis with a chloride deficiency, saline should be supplied in generous quantities. Hypodermoclyses consisting of about 80 cc. of physiologic salt solution with or without dissolved glucose should be given twice a day until hydration is complete and this point may be determined by clinical observation rather than by laboratory tests. Transfusion of blood and intravenous fluids are practically never necessary. During the days of preparation, the baby should be given 60 cc. of 5 per cent glucose by mouth every three hours since he retains this better than a formula feeding.

Immediate Preoperative Care.—Three points in the immediate preoperative care which contribute materially to the success of the operation are worth emphasizing. Open drop-ether is the anesthetic of choice since it is well tolerated by these small babies. Novocaine may be used for local infiltration or field block, but we believe that it makes the operation more difficult, not only for the patient, but also for the surgeon; and it sometimes interferes with proper wound healing. Body heat may be maintained during operation by placing a half-filled hot-water bottle beneath the baby on the operating table. It is much easier to handle the stomach during operation, and the chance of opening the duodenum is decreased, if the stomach is emptied with a tube immediately before the incision is made.

Operation.—The Fredet-Rammstedt operation is used in all cases because it is simple to execute, and it gives a permanent result. Its purpose is to relieve the obstruction at the pylorus by incising the circular muscle and spreading the cut-muscle surfaces until the mucosa completely fills the incision. Tincture of merthiolate is used to paint the skin, and a 6-cm. right rectus incision is made, starting 1 cm. below and to the right of the xiphoid cartilage. In order to deliver the pylorus into this high incision, the liver has to be retracted upward. The pylorus is delivered into the incision and the tumor held between the left index finger and the left thumb, is incised over its entire length beginning at the pyloric vein and passing upward. Only the superficial part of the tumor should be cut with the scalpel, and the incision should not be made too near the duodenal end because of the danger of perforating the mucous membrane. The cut-muscle edges are spread with a mosquito forceps beginning at the stomach end of the incision until the mucosa bulges sufficiently to completely fill the incision. The pylorus is dropped back into the abdomen and allowed to remain undisturbed while the parietal peritoneum is picked up with forceps preparatory to the closure of the abdomen. The tumor is then exposed in the abdomen to be sure that it is not bleeding. The liver, after release of the retractors, usually walls the peritoneal incision off from the rest of the abdominal cavity, thus, practically insuring against wound evisceration. To close the abdomen, we use continuous chromic in the peritoneum and anterior rectus sheath, and Michel clips in the skin. A small dry dressing is applied with adhesive straps. The clips are removed on the third or fourth day. After having tried fine silk closures, which would seem ideal for these incisions, we have

returned to the use of chromic catgut because of the great nuisance of removing the silk knots which may be quite troublesome even when the finest material is used.

Postoperative Treatment.—After operation, the baby is taken immediately to the "Pyloric Room" where the temperature is kept constant and from which all visitors are excluded. These infants in particular must be protected from all sources of infection because of their low resistance.

Each patient receives a clysis of saline and glucose once a day for the first three days after operation. The formula is increased until the baby is receiving 30 cc. every three hours at the end of 24 hours. All feedings for the first five days are given with a medicine dropper; the head of the bed is usually elevated to about 20° at feeding times, but the infant is not picked up. If the baby is to be breast fed, he is nursed once on the fifth day, twice on the sixth, *etc.*, until completely breast fed. Breast-fed babies are discharged from the hospital on the tenth postoperative day, while formula-fed babies are discharged on the fourteenth day after operation.

Complications.—It is unusual for a pyloric baby to have anything but the most uncomplicated convalescence. If the stomach has been properly emptied in the operating room and the operation correctly done, these babies do not vomit. If vomiting occurs, emptying the stomach with a tube will usually be all that is necessary. Postoperative respiratory infections are almost unheard of. One baby in this series developed an aspiration pneumonia just as he was ready for operation, but he was operated upon successfully under local anesthesia. One patient developed a duodenal obstruction from adhesions six weeks after operation, but was completely relieved by freeing the adhesions. The duodenum was inadvertently opened twice in this series, but in neither case was the convalescence affected. There has been one wound evisceration in this group but convalescence was uninterrupted after resuturing of the incision. We have had no severe wound infections and no known incisional herniae.

Healing after Fredet-Rammstedt Operation.—Dr. Martha Wollstein has made a study of healing after the Fredet-Rammstedt operation based upon a study of material from twenty-three autopsies performed from 24 hours to two years after operation. A brief summary of the important points of that study follows:

After the Fredet-Rammstedt operation, healing is brought about by cells of the serosa and submucosa, but the unstriped muscle cells take no part in the process. The incision in the pylorus is healed in nine days. The pylorus has become relaxed within two weeks. The stomach has returned to normal size within a month and the gap between the cut ends of the muscle coats has practically disappeared in six weeks. In two years only a thin line of connective tissue fibers separates these two muscle ends and the stomach is quite normal. In contrast to the operation of gastro-enterostomy which leaves the pylorus unchanged, the Fredet-Rammstedt operation cures the pylorus lesion.

Follow-up.—We have been able to follow 82 per cent of our cases. The two deaths in this series of 245 cases (1937–1946) may be briefly summarized as follows:

The baby died one month after operation from enteritis from which he was suffering at the time of operation. After operation, his vomiting ceased, but his diarrhea continued until his death. At autopsy, the Fredet-Rammstedt operation was well healed, but a severe enteritis was present, the cause of which was undetermined. He also had multiple congenital anomalies. The second baby died very suddenly 12 hours after operation. The only relevant contributing cause was a very stormy anesthetic with difficult and prolonged induction. No autopsy was obtained.

I have operated upon two patients, 29 years of age, who had had gastro-enterostomies performed at the age of six weeks for pyloric stenosis. In both patients, the pyloric tumor was present, looking exactly as it does in infancy. Microscopic study of one of these tumors showed that the only abnormal finding was hypertrophy of the circular muscle coat.

SUMMARY

1. Congenital hypertrophic pyloric stenosis occurs about seven times more often in boys than in girls.

2. Vomiting is always the first symptom and in the majority of the cases begins between the second and the fifth week of life.

3. The tumor, caused by hypertrophy of the circular muscle of the pylorus, is pathognomonic of the condition and may be felt in every case.

4. The Fredet-Rammstedt submucous pyloroplasty is the most satisfactory operation and gives a permanent result, as shown by the follow-up of the cases reported.

5. Preoperative preparation is the greatest factor in bringing the mortality to its present level.

6. The tumor disappears in about seven weeks after the Fredet-Rammstedt operation but persists throughout life in those cases who have had a posterior gastro-enterostomy done.

7. Results in 507 consecutive cases are reported.

TABLE I

No.	Deaths	Mort. %
1932—119	7	5.9%
(100)	(1)	
1937—143	0	0
1946—245	2	0.8%
—	—	—
Total 507	9	1.8%

TABLE II

M.	F.	Deaths	Mort. %
1946—206	39	2	.81%
Only Child	White	Colored	F.U.
130	241	4	82%

BIBLIOGRAPHY

Bolling, Richard W.: Congenital Hypertrophic Pyloric Stenosis. J. A. M. A., **85,** 20, July, 1925.

Donovan, Edward J.: Congenital Hypertrophic Pyloric Stenosis. ANNALS OF SURGERY, **94,** 174, November 11, 1931.

Idem.: Congenital Hypertrophic Pyloric Stenosis. J. A. M. A., **109,** 558, August 21, 1937.

Idem.: Congenital Hypertrophic Pyloric Stenosis. Am. Jour. Surgery, **39,** 377–381, February, 1938.

Idem.: Congenital Hypertrophic Pyloric Stenosis. Christopher's Textbook of Surgery, W. B. Saunders, 1941.

Downes, Wm. A.: Congenital Hypertrophic Pyloric Stenosis. J. A. M. A., **75,** 228, July 24, 1920.

Fredum, R. C., Orr, T. G., and Neff, F. C.: Congenital Hypertrophic Pyloric Stenosis. Minnesota Medicine, **20,** 508–511, August, 1937.

Knox, H. E.: Rammstedt Operation for Obstruction in Infants. Surg. Clin. of N. America, **19,** 1557–1568, 1938.

Ladd, W. E.: Congenital Hypertrophic Pyloric Stenosis. Boston Medical and Surgical Journal, **196,** 211, February 10, 1927.

Lewis, F. L. K.: Stenosis in Identical Twins. British Medical Journal, 221, February, 1944.

Robertson, D. E.: Congenital Stenosis. ANNALS OF SURGERY, **112,** 687–699, October, 1940.

Szilegyl, D. E., and McGraw, A. B.: Problems of Infantile Stenosis: With Reference to Surgical Treatment. Surgery, **13,** 764–818, May, 1943.

Walters, Waltman: Partial Gastrectomy for Hemorrhagic Gastrojejunitis following Previous Gastro-enterostomy for Congenital Pyloric Stenosis in Infancy (3 Cases). Proc. Staff Meetings, Mayo Clinic, **16,** 321–323, May 21, 1941.

Wollstein, Martha: Healing of Hypertrophic Stenosis after Fredet-Rammstedt Operation. Amer. Journal Diseases of Children, **23,** 510, June, 1922.

DISCUSSION.—DR. WILLIAM E. LADD, Boston, Mass.: I think Doctor Donovan should be congratulated on his paper and results. His ideas so closely coincide with mine that it is rather difficult to discuss his paper. Recently we have analyzed our records, and I think a report may be of interest. This analysis differs from Doctor Donovan's in the fact that it goes back many years further, and includes cases operated upon by all the surgical staff of Children's Hospital, including the resident staff, from 1915 to 1946, a period of 30 years. In that time we have had 1,145 cases. In the first 20 years, 1915 to 1935, there were 588 cases, with 35 deaths, a mortality of 5.9 per cent. In the last ten years there were 557 cases, with five deaths, a mortality of 0.8 of one per cent. In the last three and one-half years, there have been 225 consecutive cases with no deaths.

Of the five deaths in the last ten-year period, four had postmortem examinations. The causes of death were as follows: The first, aspiration pneumonia; the second was a questionable Mongolian idiot with defects of the central nervous system, kidneys and ureter; the third, pulmonary edema and terminal pneumonia. (I think this was possibly due to giving too much fluid. One should be cautious about not giving too much parenteral fluid to these small babies.) The fourth case died apparently of intestinal obstruction from congenital stenosis of the ileum which was not recognized at the time of the operation for pyloric stenosis.

I want to congratulate Doctor Donovan on the incision. We had more trouble with wounds in the first part of our series than he did. We had a few eviscerations and a certain number of cases in which the wound broke down. That may have been due to faulty technic, but I am inclined to think it was due more to the fact that in the early period we received these patients in very emaciated condition, extremely dehydrated, with extremely low resistance. We used the right rectus incision, and catgut for sutures. When we had trouble with the wounds we shifted to silk, and still had just as much trouble. More recently, we have come to the high transverse gridiron incision, and have gone back to silk. This incision gives a very good exposure and, with that and the use of silk, we have had no trouble with the wounds. I will not say that we have not had an occasional stitch that has spit-out, but not