CONGENITAL HYPERTROPHIC STENOSIS OF THE PYLORUS. WITH X-RAY PLATES.*

BY CHARLES G. MIXTER, M.D., BOSTON,

Assistant Surgeon to the Children's Hospital, Boston.

From the clinic of the Surgical Service of the Children's Hospital.

ALTHOUGH this condition was fully described by Hirshsprung in 1888 the medical profession has been slow to recognize its importance in the explanation of certain of the gastro-intestinal disturbances of infancy. This is due to the difficulties in the differential diagnosis and the failure on the part of the general practitioner to grasp the fact that here they were dealing, not with a functional disturbance but with an organic condition with a definite pathological lesion. Of late years the diagnosis is steadily being made with greater frequency, but the resort to surgical intervention is often still too long delayed.

Three factors are chiefly responsible: First, the feeling that young children and infants do not withstand severe surgical procedure well. Second, that the stenosis is a temporary condition which will right itself if the infant can be kept alive for a few months under medical treatment. Third, that following a gastro-enterostomy in infancy should the baby survive, he would develop into a sickly delicate child. If it is understood that there is present in this condition a permanent pathological lesion which no amount of medical or dietetic measures can cure, and the end results of surgical procedure can be shown to be good so far as the development of the child and the function of the gastro-intestinal tract is concerned, then it is evident that we have to deal with a surgical condition; and further, if we can show that the results of operations have been good in those cases where the condition has been early recognized and surgical intervention sought before the infant has reached the extreme stage of exhaustion and starvation, when operation has been selected as a forlorn hope, then we may expect that the medical profession and the parents will consent to operation at that early period of this disease which is most favorable to withstand operative shock, and which they have come to accept in the surgery of adults.

The pathology of this condition is simple. In gross there is found at autopsy a smooth, hard mass about the size of a pecan nut, free from adhesions occupying the pyloric end of the stomach. The mucous membrane is thickened and thrown into folds in the pars pylorica. The lumen may be completely occluded in severe types, though the degree of occlusion varies. Microscopical section shows an hyperplasia of

the circular muscle fibres and a certain amount of connective tissue increase.

The symptoms are those of pyloric obstruction and appear immediately after birth or within a few weeks. Most frequently the disease manifests itself during the first week or ten days, although the onset has been reported as late as the forty-first day. Male infants are more prone to this condition than females, the ratio being given as about eight to one. Of the ten cases at the Children's Hospital which have been proved by operation or at autopsy all were males. Vomiting is the primary and salient feature. Typically it is projectile, though occasionally there are periods of regurgitation or rarely the expulsive character may be wholly lacking. It occurs during or soon after nursing, generally within an hour, rarely two or three times in the twenty-four hours, when the obstruction is of a less degree. Water is vomited as readily as milk or other infant foods.

Visible peristalsis is marked, several waves occasionally being apparent at the same time, crossing the epigastrium from left to right. It is most noticeable with the stomach distended, but even then may only appear at intervals, if the stenosis is but partial, due possibly to a superimposed spasm. In general there is fullness across the epigastrium, particularly noticeable during peristaltic contraction and the abdomen below the umbilicus is retracted.

A constant hard movable tumor in the right hypochondrium near the median line the size of a pecan, combined with vomiting and peristalsis makes the diagnosis conclusive. Too much stress, however, has been laid heretofore on the presence of a palpable tumor, for often it is indefinite—due to its deep-seated position, and the liver may overlie it and render it impalpable. Not infrequently it is only to be felt with the aid of a general anesthetic and sometimes it cannot be demonstrated except by exploration. The absence of a pyloric tumor on palpation is never of itself sufficient to rule out hypertrophic stenosis.

The stools are usually green and slimy, containing mucus, and may resemble meconium. At other times they may approach the normal, depending on the amount of food that passes the pylorus. Constipation is the rule, though there may be frequent small dejections, of a green slimy nature. The baby nurses well, even greedily, and may cry constantly from hunger. The vomitus consists of the milk unchanged, or curdled and sour, depending on the length of time it has remained in the stomach, and may be greater in amount than the last previous feeding. There is progressive loss of weight and emaciation.

The diagnosis is difficult. Gastro-intestinal indigestion and pylorospasm are the chief confusing causes. In the first, peristalsis is absent or of very rare occurrence and the vomiting is not explosive. A proper modification of the

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milk will cure the infant. In pylorospasm the obstruction is not constant, certain of the feedings may be well borne so that several consecutive nursings or even those given for several days may be retained, and then vomiting returns and the feedings are regurgitated. The vomiting is not markedly explosive and peristalsis is absent. The amount of vomiting is not quickly influenced by substituting a proper milk formula, as in gastro-intestinal indigestion, but enough food passes through the pylorus to permit of a gradual improvement and slow gain in weight, the improvement becoming more rapid as the child grows older. There is no pyloric tumor. The vomiting finally ceases and the child develops normally.

The bismuth x-ray throws much light in those cases in which the diagnosis is difficult. In chronic gastro-intestinal indigestion, the picture is like that of the normal infantile stomach. The plate taken immediately after the feeding is given by stomach tube, shows the mass well filling the stomach and tapering down to a more or less pointed end at the pylorus with some of the bismuth already beginning to show in the small intestine. Occasionally peristaltic contractions may be observed, but they are not so well marked as in the normal adult stomach. The bismuth progresses relatively slowly from the stomach to the small intestine—in two hours about one-half of the bismuth will have left the stomach, and in four to five hours the stomach will be practically empty, which corresponds with the time required in the normal infant. There is nothing characteristic to be shown by x-ray in this condition.

In pylorospasm and stenosis, there is shown by radiogram a wide departure from the normal. There is no sharp differentiation between the two conditions, but the picture produced by pyloric obstruction, whether functional or organic, is typical. Immediately after ingestion the stomach is shown distended, the bismuth evenly distributed, and the pyloric end of the bismuth mass outlined in a characteristic prow-like rounded mass. Later bismuth will begin to appear in minute masses in the small intestine, the amount passing the pylorus depending on the degree of obstruction. The mass of bismuth which at first filled the stomach, the pyloric end being well to the right of the median line, gradually assumes a dense ball-like mass at the most dependent position of the greater curvature near the median line, though usually slightly to the left (the rapidity with which the shape is assumed depending apparently to some extent on the activity of the peristalsis). The stomach is still distended, its outline being marked by the particles of bismuth clinging to the mucous membrane, and between it and the dense central mass is a wide air space, except where the bismuth lies in contact with the greater curvature. The pyloric end maintains its rounded prow-like appearance. This condi-

tion will remain unchanged for hours, with the exception of a slight increase in the amount of bismuth in the small intestine, where the obstruction is incomplete.

The difference between the radiograph of true stenosis and that of cases which are classed as pylorospasm, is relatively minor. The plates taken immediately after the bismuth meal are identical. Later, however, when the bismuth becomes balled, the exaggerated type of peristalsis is marked in stenosis, while it is absent in spasm. The amount of bismuth passing into the small intestine depends on the degree of the obstruction, and this leads to the question, is there a distinct line drawn between these two classes of cases, each being a diagnostic entity, or do they merge from the one to the other? We know that the degree of stenosis in the organic type varies, and may we not be dealing at least in some of the cases classed as spasm, in which there is a gradual diminution in the vomiting and slow gain in weight, with a mild degree of organic stenosis, in which the passage of small amounts of food through the pylorus at first, will slowly dilate the stricture to a degree compatible with normal development. Spasm should not be excluded, but may it not be secondary to a relatively small degree of hypertrophy of the muscular fibres and stenosis of the pylorus?

Whatever the cause of the obstruction, in all those cases where dietetic measures have failed and the radiograph shows a degree of obstruction sufficient to account for a progressive loss of weight, even if the clinical picture resembles what is classed as pylorospasm, I believe that operation should be advised. For if there is obstruction and progressive emaciation under medical treatment the outcome is obvious unless the obstruction is relieved. I have been able to study four cases with the bismuth x-ray after operation.

CASE 1. No. 3516, Medical Records, Children's Hospital. Admitted March 28, 1910. W. B., male, 3½ weeks.

History. Instrumental delivery. Weight, 10 pounds. Bottle feedings. Vomiting began one week after birth. Explosive, five or six times a day. Occasionally more than at last feeding.

Examination. Well developed and fairly nourished. No palpable mass. No peristalsis. Three days later a questionable tumor was made out in the region of the pylorus.

Bismuth x-rays showed retention in the stomach, although a small amount had passed through into the small intestine 2½ hours after the meal was given.

Course. Remained in medical wards for one week, during which time various formulas were tried, but the vomiting persisted and there was a progressive loss in weight. Transferred to the Massachusetts Infants' Asylum and was given breast milk for one week without improvement. Operation by Dr. J. S. Stone revealed the usual

hard fibrous pylorus—a posterior gastro-enterostomy was done. There was very little vomiting after operation and improvement was rapid. After discharge there was some vomiting that subsided when proper milk modification was given.

Reports for observation Feb. 25, 1913. Three years old; weighs 39¾ pounds. Has been a very healthy child and has gained steadily, is being given a very liberal diet. Very occasionally has an upset from over-eating. No cathartics are required. There is no diarrhea.

Examination. Abdomen full, not distended. Scar firm, no masses or resistance felt in right upper quadrant. X-rays show the bismuth leaving the stomach with great rapidity through the gastro-enterostomy stoma, none passing through the pylorus.

CASE 2. Medical Record, Children's Hospital. Admitted June, 1912. A. G., male, five weeks.

History. Normal and well for ten days. Then vomiting began usually immediately after feedings; expulsive. Progressive loss of weight.

Examination. Emaciated. Hard mass, size of end of thumb in right upper quadrant. Peristalsis from left to right across epigastrium. X-rays show no bismuth passing through the pylorus. Transferred to surgical service.

Course. Operation, Dr. C. G. Mixer, June 22, 1912. A firm fibrous mass at the pylorus was found 5-8 inch in diameter. No opening could be detected. A posterior gastro-enterostomy was done. Vomited only twice during convalescence. A series of x-rays were taken six days after operation, showing all the bismuth passing rapidly out of the stomach by way of the enterostomy stoma.

Reports, Feb. 25, 1913. Child perfectly healthy and gaining steadily. No vomiting, bowels normal.

Examination. Scar firm, no mass or resistance in right upper quadrant. X-rays show all the bismuth leaving the stomach rapidly through the stoma.

Several attempts to pass a duodenal tube made on March 1, 1913, and controlled by x-ray show the tube passing down to the pylorus and then stopping, or in one case coiling upon itself in the stomach and then passing through the stoma.

CASE 3. Medical O. P. D. Records, No. 24,083, Children's Hospital. Seen March 19, 1912. A. D., male, six weeks.

History. Normal delivery; breast three days, then modified milk. Persistent vomiting since birth. Progressive loss of weight, visible peristalsis after nursing. No tumor palpable.

Owing to a lack of surgical beds in the hospital, the patient was referred to the Infants' Hospital.

Course. Operation, Dr. Beth Vincent, April 12, 1912. Posterior gastro-enterostomy.

Admitted to medical service, Children's Hospital, June 3, 1912.

Child healthy and gaining rapidly in weight. X-rays taken at the time show the stomach emptying rapidly through the stoma and no food passing through the pylorus.

The child continued to do well until the next winter, when during the cold weather he contracted double otitis media and later died when about eleven months old.

CASE 4. No. 4901. Medical Records, Children's Hospital. Admitted Aug. 28, 1912. A. B., male, six weeks.

History. Normal delivery, eight pounds. Breast fed; weight at end of third week, 11¼ pounds. Vomiting began at fourth week. Expulsive, during or immediately after every feeding.

Examination. Well developed and fairly nourished. Visible peristalsis, left to right after nursing. Hard, movable mass palpable at pylorus. No x-ray taken. Transferred to surgical.

Course. Operation, Dr. J. S. Stone. Posterior gastro-enterostomy. Good convalescence, very little vomiting. Rapid gain in weight. Reports for observation, April 4, 1913. Healthy, bright, lively child. Is being given a little beef juice, crackers, etc., as well as milk. No cathartics necessary. No vomiting, no intestinal disturbances.

Examination. Well developed and nourished child, scar solid, no tenderness or mass palpable.

X-ray shows an obstructed pylorus, with a rounded end to the bismuth mass and all the bismuth passing with moderate rapidity out of the stomach through the gastro-enterostomy opening.

One other case I have been able to follow, from the records of the Children's Hospital, after gastro-enterostomy, though the parents have refused to bring the child in for x-ray study. The infant has developed normally and is considered by the parents to be a healthy boy for his age, two years.

The x-ray study of cases of pyloric stenosis following posterior gastro-enterostomy is interesting. The stomach, distended before operation, soon resumes its normal size, plates taken on the sixth day following operation showing it to be within normal limits and the peristaltic contractions to be of normal intensity. In all four cases, the first plate, taken immediately after the introduction of the bismuth by stomach tube, shows the bismuth passing out into the small intestine through the stoma with marvellous rapidity. In twenty minutes the stomach is more than half empty, and in three quarters to one hour the condition is practically the same as seen in the normal stomach in four to five hours.

And naturally the query will present itself, does this condition persist, and if so will the children develop normally. The x-rays of Case 1, taken nearly three years after operation, show that the stomach empties at tremendous speed and yet the child is in all respects an unusually healthy child. In all five cases followed, the infants have developed normally and were in no wise prone to intestinal upset.

Radiographs of cases taken at varying times after operation up to three years, show that the condition remains unchanged, the pylorus remaining closed and all the bismuth passing rapidly into the small intestine through the artificial opening with the exception of what appears to be a minute amount issuing from the pylorus as it did before operation, where the obstruction was incomplete.

It is conclusive proof of the permanent character of the obstruction and further demonstrates that the rôle played by spasm in these cases of true stenosis is of minor significance.

In view of the facts demonstrated by the x-ray study of post-operative cases, the treatment of congenital pyloric stenosis is obvious. Medical treatment is clearly futile in effecting a cure, and the idea formerly held that if the infant could be tided over the first few months, the pylorus would later become patent, is also erroneous. For it has been shown, that in cases of gastro-enterostomy in the presence of a patent pylorus all the food passes into the intestine by the latter avenue and not by way of the open stoma. In this series of cases the bismuth meal entered the small intestine through the artificial opening. In an attempt to pass a duodenal catheter, the tube traversed the greater curvature of the stomach, passed the stoma to the obstructed pylorus, where it was deflected upward and made a complete circuit of the viscus and finally entered the small intestine through the gastro-enterostomy opening.

Surgery offers the only curative measure and should be resorted to at the very earliest moment the diagnosis can be made. The bismuth x-ray is a conclusive means of determining the presence of obstruction at an early period and where the diagnosis is in doubt. The Loretta pyloroplasty and gastro-enterostomy are the operations applicable to this condition, and the latter is generally conceded the procedure of choice, as it is the most rapid and simple, and there are no untoward remote after-effects.

CONCLUSIONS.

In conclusion I would like to emphasize the following points:—

1. An early diagnosis is the most essential factor in lowering the mortality from this disease.
2. The bismuth x-ray will conclusively demonstrate the presence of pyloric obstruction.
3. In any case in which careful medical treatment has been unable to check the vomiting and progressive loss of weight, and where a pyloric obstruction can be demonstrated by x-ray, even if the differential diagnosis cannot be definitely made between hypertrophic steno-

sis and pylorospasm, exploration should be advised.

4. X-rays, taken at varying intervals after operation, show the pyloric obstruction to be a permanent condition.

5. In the cases studied, spasm was of no importance as a factor in the obstruction.

6. In true stenosis, therefore, surgery offers the only hope of cure.

7. The infant develops normally after posterior gastro-enterostomy.

I wish to express my thanks to the medical staff for their kindness in permitting me to have access to their records; to Dr. Maynard Ladd for the photographs of gastro-intestinal indigestion and pylorospasm, by whose kind permission they are published, and to the x-ray department for their coöperation in obtaining the radiograms.

LEGENDS FOR X-RAY PLATES.

Series 1. Gastro-Intestinal Indigestion. Plate 1. Immediately after bismuth meal. Stomach of normal size—pyloric end of bismuth mass is not rounded. Small amount of bismuth in small intestine.

Series 1. Plate 2. Two hours after bismuth meal. Stomach more than one-half emptied—bismuth in considerable quantity in small intestine—no "balling" of bismuth in stomach.

Series 2. Pylorospasm. Plate 1. Immediately after bismuth meal. Stomach distended—rounded end of bismuth mass—none in small intestine.

Series 2. Plate 2. Two hours after meal. Balling of bismuth in dependent portion of stomach. Air space between wall of stomach and mass—small amount in intestine. Absence of peristalsis in stomach.

Series 2. Plate 3. Three and one-half hours after bismuth. Immediately after next regular feeding, stomach distended above bismuth mass. No peristalsis. Small amount of bismuth in intestine.

Series 3. Hypertrophic pyloric stenosis. Before operation—A. G. Plate 1. Immediately after bismuth meal. Stomach distended—typical prow-like end of bismuth mass at pylorus. Well-marked peristaltic contractions—no bismuth in intestine.

Series 3. Plate 2. Ten minutes after bismuth meal. Rounded pyloric end of stomach—marked peristalsis—as yet no balling. No bismuth in intestine.

Series 3. Plate 3. Two hours after bismuth. Some balling of bismuth. Large air-space. Small amount of bismuth in intestine.

Series 3. Plate 4. One-half hour after bismuth. A. M. Rounded pyloric end. Beginning balling. Peristaltic contractions. Bismuth in median line.

Series 4. Hypertrophic pyloric stenosis—before operation, J. McK. Plate 2. One hour and ten minutes after meal. Strong peristaltic contractions. Bismuth to left of median line.

Series 4. Plate 3. Three hours and thirty minutes after meal. Outline of stomach shown by bismuth clinging to mucous membrane.

Series 4. Plate 4. Five hours after meal. Marked balling—large air-space—no bismuth in intestine.

Series 5. Hypertrophic pyloric stenosis—after operation—6 days. A. G. Plate 1. Ten minutes after bismuth. Stomach normal in size. Rounded pyloric end of bismuth. Bismuth passing rapidly into intestine through stoma.

Series 5. Plate 2. Twenty minutes after bismuth. Large amount of bismuth in the intestine—stomach contracting well and at least one-half empty. Slight amount of bismuth apparently passing through pylorus.

Series 5. Plate 3. Three quarters of an hour after bismuth. Stomach emptying rapidly and contracting well. Fine line of bismuth passing through pylorus.

Series 6. Hypertrophic pyloric stenosis. After operation, 6 months. A. G. Plate 1. Immediately after bismuth. Stomach emptying rapidly through stoma.

SERIES 1. PLATE 1.

SERIES 1. PLATE 2.

SERIES 2. PLATE 1.

SERIES 2. PLATE 2.

SERIES 2. PLATE 3.

SERIES 3. PLATE 1.

SERIES 3. PLATE 2.

SERIES 3. PLATE 3.

SERIES 3. PLATE 4.

SERIES 4. PLATE 2.

SERIES 4. PLATE 3.

SERIES 4. PLATE 4.

SERIES 5. PLATE 1.

SERIES 5. PLATE 2.

SERIES 5. PLATE 3.

SERIES 6. PLATE 1.

SERIES 6. PLATE 2.

SERIES 6. PLATE 3.

SERIES 7. PLATE 1.

SERIES 7. PLATE 2.

SERIES 7. PLATE 3.

SERIES 8. PLATE 1.

SERIES 8. PLATE 2.

SERIES 8. PLATE 3.

SERIES 8. PLATE 4.

SERIES 8. PLATE 5.

SERIES 9. PLATE 1.

SERIES 9. PLATE 2.

SERIES 9. PLATE 3.

Series 6. Plate 2. One-half hour after feeding. Rounded end of bismuth mass at pylorus.

Series 6. Plate 3. Stomach contracting and discharging contents through stoma. None passing through pylorus.

Series 7. Hypertrophic pyloric stenosis. After operation—nine months—A. G. Plate 1. Immediately after feeding. Condition the same as in previous series.

Series 7. Plate 2. Fifteen minutes after bismuth. No bismuth passing through pylorus.

Series 7. Plate 3. One hour after bismuth. Stomach emptying through gastro-enterostomy opening.

Series 8. Hypertrophic pyloric stenosis. After operation—seven and one-half months. Plate 1. Immediately after feeding. Stomach emptying rapidly—pylorus rounded.

Series 8. Plate 2. Fifteen minutes after feeding. Condition the same—more bismuth in intestine.

Series 8. Plate 3. One hour after bismuth. Probably one-half bismuth in intestine.

Series 8. Plate 4. One and one-half hours after bismuth.

Series 8. Plate 5. Two hours and fifteen minutes after bismuth. Stomach perhaps three-fourths empty—contracted.

Series 9. Hypertrophic pyloric stenosis. Plate 1. A McJ. Two months after operation. Twenty minutes after bismuth. Rounded pylorus—bismuth passing through stoma.

Series 9. Plate 2. W. B., 3 years after operation—15 minutes after feeding. Child allowed to drink bismuth instead of using stomach tube. Empties with extreme rapidity.

Series 9. Plate 3. A. G., nine months after operation. Duodenal catheter has passed to the closed pylorus where it has been deflected up to the cardia and then down and finally out through the gastro-enterostomy opening.

Clinical Department.

REPORT OF THREE CASES OF ACUTE ANGULATION OF THE TERMINAL ILEUM CAUSING INTESTINAL OBSTRUCTION, FOLLOWING OPERATION FOR AND DRAINAGE OF ACUTE SUPPURATIVE APPENDICITIS.

BY C. A. ROEDER, M.D., GRAND ISLAND, NEBRASKA,

Surgeon to St. Francis Hospital.

PROMPTED by Cheever's article on the above subject (BOSTON MEDICAL AND SURGICAL JOURNAL, May 15, 1913), I am led to report three cases similar to his. It must be a relatively frequent complication following operations for the pus appendix, and since Cheever has called our attention to it, I am sure, directly or indirectly, we shall hear a great deal more of it. I have three cases to report, of two of whom a history and record have been kept. The remaining one I have only a mental record of, which, however, will serve the purpose. All three cases occurred in men; all had pus and were drained, two with drains extending to the bottom of the pelvis, and all recovered. In each case at the second operation the terminal ileum, varying from six to ten inches, was bound down to the pelvic floor.

We hesitate to attack surgically an acute area

such as will be found in these cases four weeks or more after the operation. This led me to short circuit the ileo-cecal region by an ileo-sigmoidostomy, working in a clean field and draining the obstructed ileal contents into the sigmoid, a portion of the intestine capable of very little absorption and whose contents are easily evacuated.

Whether or not the pus, the drainage tube, the distention, the ascending position of the terminal ileum, or the Fowler position may cause this acute obstructive angulation near the end of the small bowel, we know it occurs and must be treated promptly and judiciously, because it is at this time that any procedure determines the fate of the patient.

In all three of our cases we opened the abdomen as soon as the obstruction was evidently not yielding to medicinal measures such as lavage, purgatives hypodermatically, enemata, and hot applications, not allowing more than twelve hours to pass from the beginning of the first symptom before performing celiotomy. We used the Kameron incision through the left rectus, the ileum approaching the obstruction was joined to the sigmoid as low as possible with a medium-sized Murphy button, and the abdomen quickly closed. This gives immediate drainage, as shown by one patient evacuating the contents of the obstructed ileum before being removed from the operating table. The time consumed is short, about ten to fifteen minutes, the patient lightly under ether or gas and protected by nerve block. In our experience it has not been necessary to operate later to relieve the restricted ileum. In one case a cecal fistula, which had stopped draining owing to the ileal kink, opened up four days following the anastomosis.

CASE 1. Male, age 17. Was seen at home in the country on Jan. 15, 1913, complaining of general abdominal pain, more marked in the right lower quadrant. Had been sick for three days with symptoms of acute appendicitis and presented a typical picture of peritonitis following such a condition. Abdomen opened through Kameron incision, free sero-purulent dark brown pus flowed out. Appendix deeply buried, and owing to great distention cecum tapped with a No. 24 catheter sewed in similar to a Senn gastrostomy. Tube placed at bottom of the abscess cavity where appendix lay, which was left in place two weeks. The temperature gradually dropped to normal and remained so until the twenty-ninth day, when septic temperature curve started. Brought to hospital, abdomen opened and pocket of pus behind cecum evacuated and drained. Temperature normal in ten days with wound draining. Started a septic curve again on seventeenth day following last operation. Abdomen opened, attempted to remove appendix, which was impossible, closed fecal fistula and placed drain to bottom of pelvis. Fistula remained closed, but temperature above normal in the evening for four weeks. Abdomen again opened and remaining stump of appendix removed, drain placed in cavity occupied by stump of appendix.