CONGENITAL ATRESIA OF THE ESOPHAGUS, WITH REPORT OF THREE CASES

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Congenital atresia of the esophagus is everywhere considered one of the rare anomalies. Sir Morell Mackenzie¹ (1884), after an exhaustive search could collect only sixty-two cases from the literature, and added only one of his own. Shukowsky,² with a hospital experience of 50,000 new-born babies, saw only one case. And yet within a period of one year three of these cases came under my observation, and I have knowledge of at least one other within the following year.³ Even more striking was Hirschsprung's⁴ experience, who, in 1861, in Copenhagen, with only 180,000 inhabitants, saw four of these cases within a period of seven months. Although such grouping is a matter of chance, yet one cannot help but wonder whether these cases are not more frequent than we have been led to think.

The cases here reported were all under exact observation in hospitals, and in each instance a complete autopsy was obtained.

While many different kinds of congenital anomalies of the esophagus are described, one type is the rule (70 + per cent.). This is the so-called "inosculating type," in which the upper end of the esophagus ends in a free dilated pouch, while the lower end passes from the stomach into the trachea (or, very rarely, into a bronchus). This predominance of one type suggests a uniform embryologic origin. This interesting feature will be discussed fully elsewhere by Professor Zeit,⁵ and I refer anyone interested in that phase of the subject to his paper. My own report will be chiefly clinical, together with some theoretical considerations that naturally suggest themselves.

The cases that I have seen have all been, as were Hirschsprung's,⁴ of the predominating type. So distinctive and uniform were the clinical features in all of them that it was easy to venture an exact anatomic

1. Mackenzie, Sir Morell: *A Manual of Diseases of the Throat and Nose*, 1884.

2. Shukowsky and Baron: *Archiv für Kinderheilkunde*, Bd. 58, 1912, pp. 191-8.

3. Since writing the above John Lovett Morse, in the *Archives of Pediatrics*, July, 1912, has reported having seen three cases in the past year, with one autopsy.

4. Hirschsprung: *Den Medfødte Tillerkning af Spiseroret*. Copenhagen, 1861. Also *Hospitalstidende*, 1895, No. 42.

5. Zeit, F. R.: *Med. Research*, 1912, xxvii, No. 1.

diagnosis, *ante mortem*, as was done in the second and third babies, after having followed the first. A detailed description will be given of the first case only, the others so closely resembling it that only the differences will be pointed out.

Fig. 1.

Fig. 2.

Fig. 1.—Congenital atresia of the esophagus, posterior view. Case 1. The dilated upper end of the esophagus is shown extending from below the rima glottidis to the bifurcation of the trachea; the narrower lower portion from the stomach into the trachea at the bifurcation. Drawn by Miss E. Von Stoesser.

Fig. 2.—Same as Fig. 1, anterior view. Upper dilated end of the esophagus partly hidden by the thyroid gland and the trachea. The lower portion of the trachea and the bronchi are laid open showing a sound (black) passing from the stomach through the lower portion of the esophagus into the trachea at the bifurcation. Drawn by Miss E. Von Stoesser.

CASE REPORTS

CASE 1.—Baby O., colored, born at full term in St. Luke's Hospital Feb. 6, 1910, on Dr. Paddock's service. Family history negative. Birth-weight 5 pounds 9 ounces; length 45 cm. Outside of the fact that it had a very low temperature (92 F.) a few hours after birth the baby attracted no special attention until it began to expel mucus from the mouth. Four hours after birth the nurse recorded "Spitting up mucus"; two hours later, "Mucus drawn from throat by intern." On the following day she recorded: "Mucus almost continually coming from nose"; and following a teaspoonful of water, "Water coming back through the nose." On the third day she recorded (baby taken) "to mother; almost strangled; milk came back through nose." Because of the complete inability to retain anything the child was transferred to the pediatric service on the third day.

The baby was manifestly in an alarming condition, was emaciated, had lost nearly a pound, the skin was dry and the tissues were beginning to feel hard; the eyes were sunken—in short there was the typical appearance of a baby long deprived of water. From the lower angle of the mouth there flowed a constant stream of stringy mucus and saliva, running over and excoriating the cheek and chin. In front of each nostril there was a considerable quantity of white, frothy discharge that bubbled to and fro with each respiration. When the child was nursed or given water from a bottle it swallowed greedily, but after one swallow the milk or water came back through the mouth, and in jets from both nostrils, synchronous with each act of swallowing. Then with the first breath it would suddenly stop nursing, would cough, become deeply cyanotic, and look as if it were choking to death. After expelling the water by coughing, and especially if the baby was inverted, it would gradually return to normal. *The upper part of the abdomen was somewhat distended, the lower part rather flat.*

A rubber catheter inserted through the mouth and always ending abruptly at a distance of about 12 cm. from the lips, clinched the diagnosis. The normal distance to the cardiac end of the stomach is about 17 cm. in the new-born.

Mouth feeding was discontinued after the diagnosis was made and normal salt solution and peptonized milk were given by rectum. On the fifth day the baby was operated on by Dr. L. L. McArthur. A jejunostomy was performed and a catheter left in place for feeding. Although the operation was well borne the general condition grew steadily worse; the baby withered and dried up perceptibly from day to day, the respirations could scarcely be seen, the pulse became imperceptible, the tissues everywhere became dry and wood-like. The baby for several days resembled an automaton rather than a living thing and died on the tenth day—a typical picture of water starvation in an infant.

On the fourth day the temperature went up to 100.2 F. and reached nearly that point every day, except the last, when it became subnormal.

The weight dropped from 5 pounds 9 ounces to 4 pounds 9 ounces on the third day and to 4 pounds and 3 ounces on the last day.

Necropsy.—This was performed by Dr. E. R. Le Count, to whom I am indebted for the beautifully dissected specimen shown in the illustration.

The upper end of the esophagus ended bluntly 4 cm. below the rima glottidis, and 1 cm. above the bifurcation of the trachea. It was considerably dilated, of a practically uniform diameter of 1 cm. throughout.

The lower end of the esophagus was 3 cm. long and passed directly from the normal cardiac end of the stomach into the posterior portion of the trachea just at the bifurcation. The gastric end was 5 mm. in diameter, and the tracheal end 4 mm. There was no connection whatever between the two portions of the esophagus.

Further anomalies were found in the microscopical anatomy of the spinal cord, and in the ureters. On each side there were two ureters, one normal and the other running parallel to it, and opening into the bladder by an opening close to

that of the normal ureter but distinctly separate from it. Both supernumerary ureters tapered off toward the renal extremity and ended freely, the right 4.5 cm. and the left 8.5 cm. long.

The lungs showed a bronchopneumonia.

CASE 2.—Baby N. The baby was sent to the pediatric service at Wesley Hospital from the out-patient department of the Lying-in Hospital when 3 days old, with a probable diagnosis, for obvious reasons, of bronchopneumonia. There was some fever, the breathing was rapid, both lungs were covered with moist râles, and there were spells of alarming cyanosis. The baby had retained nothing since birth.

The history of vomiting, and of spells of cyanosis, the general appearance of the child, its striking resemblance to the former case, the withered, dried-up, mask-like face and sunken fontanel, the constant flow of saliva from the mouth, reddening the chin and cheek, but, above all, the frothy, bubbly, white discharge before the nostrils that could hardly come from anything else, at once suggested the diagnosis of an atresia of the esophagus. The catheter stopping abruptly, as in the former case, at 12 cm. from the lips, fixed the diagnosis as to the upper end of the esophagus. That the lower portion of the esophagus formed a communication between the stomach and the respiratory tract was evident from the fact that the stomach was distinctly distended and tympanitic while the rest of the abdomen was slightly flattened.

On the fifth day of life a gastrostomy was performed by Dr. H. M. Richter, leaving a catheter *in situ* through which the baby was fed on breast milk. When the tube was clamped after the food was introduced, the child would soon cough, breath 100 to 150 times a minute, become deeply cyanosed, and would seem to be literally choking to death. If the tube was now opened to prevent the child from dying, the milk would well out through it, often in spurts as the child coughed. A colored fluid introduced into the stomach in this manner was recovered in part from the mouth. So alarming were the symptoms of drowning in this instance that it was not repeated. When the stomach was empty and the tube open there was an audible to and fro movement of air through the tube.

Necropsy.—This was performed by Dr. F. R. Zeit and the specimen forms the basis of the paper on this subject by Dr. Zeit.⁵ The findings differed only in unimportant details from those of the first case. The upper sack ended blindly 1 cm. above the bifurcation of the trachea; was 4.5 cm. long, and was even more dilated (1.7 cm.) than in Case 1. The lower portion was 7.5 cm. long, and tapered from a diameter of 7 mm. at the opening into the stomach to 4.5 mm. at the opening into the mid-posterior point of the bifurcation of the trachea. There was again no connection whatever between the two portions of the esophagus. No other anomaly was found.

The lungs showed a bronchopneumonia.

CASE 3.—Baby W., born at Hotel Dieu, in the private service of Dr. G. J. Hagens, to whom I wish to express my indebtedness for permitting me to see the case together with Dr. J. H. Hess, and Dr. H. M. Richter, and also for allowing me to report it with the other two. This child had all the essential symptoms and findings of the other two and detailed description would be only repetition.

The diagnosis was established on the third day by the inability to pass the catheter into the esophagus more than about 12 cm. from the lips. It was decided that an operation was not indicated. The baby died on the eighth day. The autopsy performed by Dr. Hess showed identically the same lesion as the other two. The upper end of the esophagus was 4 cm. long, 1 cm. wide, and ended 1 cm. above the bifurcation of the trachea. The lower portion was 6 cm. long, 0.7 cm. in diameter near the stomach and slightly less at the other end. In this case the tracheal end of the lower portion of the esophagus emptied into the posterior median line of the trachea 1.5 cm. above the bifurcation.

The only other anomaly found was a single horse-shoe kidney.

Out of sixty-two cases of congenital malformation of the esophagus reported by Mackenzie,¹ forty were of the "inosculating type," such as the three here reported, and in three others the lower portion of the esophagus connected with a bronchus (70 per cent. in all). In nine cases there was a simple blind termination; in two a normal esophagus except for a fistulous opening into the trachea, and in three others there was a membranous obstruction. In five cases there was a complete absence of esophagus, but this occurred only in extreme general malformations (acardiacus). Thus about 75 per cent. of all cases in which there is any esophagus at all belong to the type here reported.

In one-third (Kraus) to one-half (Happich⁶) of these cases other anomalies are present, beside that of the esophagus.

We have in these cases of atresia of the esophagus new-born babes that are deprived of food, and what is far more vital, of water, and it is of interest to go a little deeper into the symptomatology of this almost unique condition.

Birth-Weight.—It is commonly stated that these babies are born small and weak. Ahlfeld⁷ has attributed this to the fact that they do not swallow amniotic fluid as the normal fetus does, and, deduced from this, that the amniotic fluid helps to nourish the fetus. Schwalbe⁸ does not agree with him, his own two cases being well developed and of normal weight. My cases weighed, respectively, 5 pounds, 9 ounces; 7 pounds, 12 ounces; and 7 pounds; an average of nearly 7 pounds. Ahlfeld's view seems further untenable in the "inosculating type," because we have here a passage from the mouth to the stomach by way of the trachea that is practically as patulous as the normal esophagus. It is hard to see why swallowed amniotic fluid should not find its way to the stomach by this route.

Length of Life.—All of the cases of the type here reported die. Mackenzie¹ reports twenty-five deaths out of thirty-seven in the first week. These of the present series lived, respectively, 9, 7½ and 8 days, under very different conditions that do not seem to have altered materially the course. Thus the first baby had a major operation and received about 30 ounces of fluids altogether, chiefly by tube into the jejunum and a small amount by rectum; the second, too, was operated and throughout its life most nearly approached absolute starvation, receiving only two saline enemas, and the rest of its fluids, water and milk, by tube into a stomach that expelled practically everything immediately; the third did not undergo an operation, and received about 2 quarts of normal salt solution by hypodermoclysis during the last four

6. Happich: Inaug. Dissert., Marburg, 1905.

7. Ahlfeld: (a) Lehrbuch der Geburtshelkunde. (b) Missbildungen.

8. Schwalbe: Missbildungen.

days of its life, and yet lived a day less than the less well developed first baby with its more serious handicap.

Cause of Death.—A bronchopneumonia, as in these cases, usually terminates the picture because of the favorable conditions for the aspiration of food into the lungs. It is hard to tell how great a part this plays, as compared with pure water starvation in bringing about the fatal end. The clinical picture is that of starvation, of desiccation. The fairly uniform length of life would lead one to think that the figures so obtained nearly represent the length of time a new-born babe can live without water. While death from strangulation often seems imminent, it apparently is rarely the cause of death, although Porro reported one such death. Whether the baby is fed by mouth or not is manifestly a large factor in determining whether it shall strangle to death, or die of a terminal pneumonia; or of simple starvation.

Loss in Weight.—This is rapid and persistent but especially so in the first few days when the tissues are still filled with water. Thus these three infants that weighed 5 pounds, 9 ounces; 7 pounds, 12 ounces; and 7 pounds, lost, respectively, 1 pound, $1\frac{1}{2}$ pounds and 2 pounds in the first three days; and a total of 1 pound, 6 ounces; 2 pounds, 6 ounces; and 2 pounds, 14 ounces. The total loss in weight was thus, respectively, about 25, 30 and 40 per cent., with nearly 75 per cent. of the loss in the first three days.

The Temperature in each case showed a distinct elevation after the second or third day, a phenomenon that is by no means rare in the new-born. This so-called "inanition temperature" of the new-born lasted in the first case from the fourth to the seventh day with 100.4 F. as a maximum. In Case 2 the temperature rose gradually with wide remissions from the third day to 102 F. on the last day. In Case 3 the temperature was slightly above normal the first two days, went to 102 F. on the fourth day and remained between 100 and 101.6 F. to the end. The rise in temperature toward the end was probably due to the terminal pneumonia. In Case 1, however, with a pronounced bronchopneumonia, the temperature was almost constantly subnormal during the last three days, a condition that is quite often seen with a terminal pneumonia in marantic babies.

Sclerema.—In each of these cases the skin became dry, leathery and non-elastic, and toward the end the subcutaneous tissue everywhere was board-like, so that the bones could not be felt as structures separate from the overlying tissues. The face became almost immovable, expressionless and mask-like. This condition is doubtless due to water starvation, and probably a solidification of the subcutaneous fat takes place.

The *meconium* did not differ in appearance from that passed by the normal new-born baby. It was possibly slightly scantier, and in Cases 1

and 3 was passed only with the aid of an enema, or suppository. In Case 2 there were two to four meconium stools a day for several days after the baby entered the hospital.

Attacks of Suffocation and Cyanosis occur in all of these cases depending in number and severity on whether the child is given food, or water, by mouth. They are evidently due to the aspiration into the lungs of the fluids that the child is attempting to swallow and that fill the mouth, pharynx and nasal passages. These attacks can likewise occur, in the "inosculating" cases, by flooding of the lungs with fluids from the stomach in the act of vomiting as was well shown in Case 2.

A most characteristic symptom is the constant flow of saliva from the mouth and the presence of a frothy discharge in front of the nostrils. The saliva that is normally secreted in considerable amount cannot be swallowed in these cases and must flow from the mouth. Regurgitation through the nose occurs because the mouth and nasopharynx are filled with fluid while the mouth is closed in the act of sucking. The nose is the only outlet. Both of these symptoms can hardly come from any other condition in the new-born.

Abdomen.—It is commonly stated that babies with congenital atresia of the esophagus have a sunken abdomen. This is only partly true. In each of these cases the epigastric region was distinctly bulging and tympanitic, while the lower part of the abdomen was flattened. This condition is manifestly due to the fact that air passes into the stomach from the trachea through the lower part of the esophagus, but not beyond the stomach. That this does happen was shown in the second case in which there was audible respiration through the catheter inserted into the stomach through the abdominal opening.

Diagnosis.—The diagnosis is easily made from the following symptoms:

1. Characteristic return of swallowed fluid from the mouth and through the nose in jets, synchronous with the act of swallowing.
2. The constant flow of saliva from the mouth and the presence of a frothy secretion before the nose.
3. Alarming attacks of suffocation and cyanosis with each attempt at mouth-feeding.
4. The attempt to pass a sound establishes the diagnosis and also the point of obstruction. The normal distance from the lips to the cardiac end of the stomach is 17 cm. in the new-born, and the minimum diameter of the esophagus 4 mm. (Mackenzie¹).
5. The "inosculating" type can be diagnosticated when the stomach is found distended with air.

Treatment.—No treatment so far has been of any avail. To attempt to feed such an infant by mouth only hastens the fatal end. Rectal

feeding in the new-born can be only a very temporary measure. A radical operation that would aim at a *restitutio ad integrum* is hardly to be considered in the new-born and not much more so in the older child if it should survive. Hoffmann attempted it, but abandoned it for a gastrostomy.

Temporizing operations have accomplished nothing because of the practical impossibility of nourishing the child under the conditions imposed. Gastrostomy, recommended by Sedillot, has been performed a number of times (Steel, Hoffmann, Happich, Villemin, Kirmisson, Dickie, Demoulin,⁹ etc.). In every case, as in Case 2, the result has been the same, the fluid introduced into the stomach has flooded the lungs, and invited death by suffocation and bronchopneumonia. It is clearly contra-indicated in cases in which a communication between the stomach and trachea has been diagnosed, and these probably constitute 80 to 90 per cent of all cases of atresia. Surgical closure of the cardiac end of the esophagus, if possible, would make this operation more rational, but would make any later radical operation still more formidable. In cases in which there is a simple blind termination of the upper end, with no lower end communicating with the trachea, simple gastrostomy would be the operation of choice.

Jejunostomy, recommended by Demoulin,⁹ and performed in Case 1, would seem sounder theoretically, but it is very doubtful if a new-born infant, thus handicapped, ever could be nourished sufficiently by jejunal feeding to more than simply prolong its life a few days, to say nothing of building it up for a future radical operation. These cases are unfortunately rarely diagnosed before so enormous a loss in weight has taken place that this alone jeopardizes the child's life. Even if a child could be fed in this manner it would be doomed to perpetual feeding through a tube, and to other embarrassments that would make life unbearable; or else it would have to submit later to a radical operation that would offer little if, indeed, any hope. While the utter hopelessness of these cases, if untreated, justifies surgical procedures that would otherwise seem too daring; nevertheless the physician who, after making his diagnosis of congenital atresia of the esophagus, decides to let his little patient die undisturbed, as in Case 3, can amply justify his course.

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9. Demoulin: Bulletin Société de Chirurgie, July 5, 1904.