<http://www1.umn.edu/eatef/index.html>

a. Overall goals

Our late results, so far, have been excellent. All children, even those who began with the longest gaps or had failed previous operations, have had a successful true primary repair. Those who are far enough out from the repair (usually a year), to allow complete resolution of anastomotic strictures and to have overcome oral aversion, are eating normally. One child came to us with a tracheostomy in place and although he now has a very satisfactory esophagus, he will not learn to eat until the tracheostomy can be removed. We believe this will be accomplished this summer. Oral aversion seems to be the most lingering barrier to eating a normal diet. Although oral aversion may require a great deal of effort by the parents, we believe that in all cases it will be solved.

As the testimony to how well they eat, we published our long term follow-up studies and found the weight of our patients was within the normal range. In fact, their weights slightly exceeded their range of heights ([reference 5](http://www1.umn.edu/eatef/references.html)). The size distributions were almost precisely normal indicating good nutrition and growth and development.

All children were eating whatever they wished and, at the time the study was completed, no child two years after repair, still had a gastrostomy tube ([reference 5](http://www1.umn.edu/eatef/references.html)). With the referral here of increasingly complicated cases of EA in children who arrive at a year of age or older, not all g-tubes have been out by the age of two.

b. Esophageal Function

The normal esophagus empties in to the stomach by a combination of muscular contraction (peristalsis) and gravity. A rhythmic wave of peristalsis which is propagated within the muscle itself, begins in the throat and passes down to the stomach. It has been believed that the vagus nerve provides the stimulus for the contraction wave. But, both older experimental studies as well as our clinical observations indicate that the contraction stimulus passes within the muscle itself. Unfortunately, the contraction impulse does not pass across a repair site. Consequently, wherever a primary repair has been done or a segment of esophagus removed and the ends joined together, the contraction wave will not pass smoothly to the lower portion of the esophagus.

The esophageal muscle below the repair site does have an intrinsic firing mechanism, however, and contractions will occur. The contractions they are not smoothly rhythmic and progressive. As a result, any child who has had an EA repair, are considered to have disordered function of the lower esophagus. The practical consequences, however, do not appear to be great. The contractions will empty the esophagus satisfactorily into the stomach and the children are able to eat whatever they wish. A normal diet can be enjoyed.

On the other hand, interpositions of colon and stomach have no peristalsis and can empty only by gravity. Therefore, these children may have significant eating difficulties and may not be able to eat whatever they wish or enjoy a normal diet.

c. GE reflux

In our opinion, follow-up evaluations should be done for GE reflux. Reflux is common, particularly after very long gap repairs. In our patients, fundoplications have been done in about 30% of the children and more often in the long gap patients. We recommend a fundoplication for significant GE reflux, because of the uncertainties of treatment by medications alone ([reference 6](http://www1.umn.edu/eatef/references.html)). Whether or not fundoplication will eliminate the need for long term antacid medication remains to be seen. We still recommend, therefore, esophagoscopy and biopsies every 1-3 years to make sure significant inflammation of the esophagus has not developed. Because our final goal, which has been realized in a large majority of our patients, is to be medication free, this follow-up is necessary. More time will be needed to accurately answer how often the potential problems of GE reflux occur, particularly in the children who have had very long gap repairs.

d. The very long term

For the very long term, we do not foresee any problems with the esophagus after a true primary repair. There are patients who had a relatively straight forward repair of EA/TEF forty years ago. The function of their esophagus appears to be very satisfactory. This should be true even for the longest gap repairs, because of the great growth potential inherent in the esophagus of all babies. It should not matter that originally the esophageal pouches were relatively short. The growth potential should more than compensate for this beginning.

In summary, we have shown that true primary repair is possible, apparently in all EA patients. This esophageal repair will reliably allow the children to eat normally and not be dependent on a g-tube. We believe the benefits of this approach will only increase with time. There seems to be no evidence for late problems arising with the esophagus, however, the potential for difficulties from untreated GE reflux remain. Clearly, however, after a few years, even the very long gap repairs cannot be distinguished from normal by what they eat.

The issues before esophageal atresia repair:

The baby's condition before these large operations is important. But in addition to the baby's general condition, there are at least three important issues which will affect the treatment plan. These are whether or not the baby: (1) is very premature, (2) or has other serious birth defects in the heart, kidney or elsewhere or (3) has a type of esophageal atresia that will be difficult to repair. These are potentially complicating issues that may significantly affect the proposed operation in terms of either timing or the final result.

a. Other birth defects

The presence of other birth defects, such as in the heart, chest, abdomen, head or limbs may be very important to the baby. The tendency of other birth defects to occur with EA/TEF is well known and termed the VATER or VACTERL associations. These letters stand for such defects as: missing or deformed Vertebrae, absence of an Anus, abnormal kidneys (Renal), malformed forearm bones (Radius or Limb), or blockages in other parts of the intestinal tract as well as heart (Cardiac) and brain lesions. The TE is from tracheoesophageal anomalies.

There are so many possibilities for birth defects other than EA/TEF, that a discussion of them will not be presented here. The presence and consequences of these defects can better be discussed by the baby's doctors.

b. Prematurity

Some EA/TEF infants are born very prematurely. If the baby is very small and underdeveloped or has very immature lungs, the surgical repair of the EA/TEF may be delayed until the situation is much improved. Treatment by the neonatologists has greatly improved, nevertheless, prematurity influences the treatment plan. Among the possibilities, are waiting until the child is larger and vigorous enough for the operation. In general, a weight of 1,700 grams or about 3¾ pounds would be preferable.

If the wait for repair will be prolonged, or the lungs are being injured by acid coming up from the stomach through the TE fistula, then an initial operation may be recommended. A gastrostomy and/or tying off of the fistula (TEF) may improve the situation until the primary esophageal repair. Again, prematurity and its consequences for the EA repair are best left for the health care providers to discuss.

c. The gap length - the barrier to true primary esophageal repair.

Once the above issues have been resolved, the child would be ready for esophageal repair. Currently, by far the most important issue affecting the repair itself is the distance or gap between the two esophageal segments. A primary repair consists of sewing the two esophageal segments together to form a tube. This is the simplest and best method of repair, but if the ends are too far apart it may not be possible.

As shown in Figure 1, the gap between the esophageal ends is longest in Types a and b. This is the most difficult end of the EA spectrum to repair. Usually, the gaps in Type c and d are short enough to allow a straightforward primary repair. But even in these groups, some infants may be considered by the surgeon to have too long a gap. As a result, even some babies with Type c EA/TEF, are also repaired with a substitution graft.

The distance is usually measured by an x-ray before the surgical repair. The gap length is expressed in terms of centimeters (cms) or vertebral bodies (Figures 2,3,4). Vertebral bodies grow with the child, so if a gap is about five vertebral bodies long, the distance in cms will depend on the size and age of the child. In a small baby,a vertebral body would be less than a cm, and a four vertebral body gap might be about 3 cm long. At about 6 months of age, however, the distance would be about one cm per vertebral body. As the child grows larger a vertebral body would be greater than one cm. The distance given in vertebral bodies gives an idea of the relative amount of esophagus missing. For example, in Figure 3, virtually all of the intrathoracic esophagus is missing: a gap of about eight vertebral bodies but, the gap is only about 5 cm long. Figure 4, a gapogram of a larger child, shows a gap of 6.2 cm but a space of only about six vertebral bodies. The absolute distance (in cm) will also indicate the difficulty with a primary repair.

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| Figure 2:  http://www1.umn.edu/eatef/images/fig2.gifAn x-ray study which allows the gap length between the two esophageal segments to be determined. The upper esophageal pouch is outlined by the black arrows. There is no TEF and this is an example of pure EA (type a). A contrast agent (usually barium) has been put into the stomach through the g-tube just prior to making this x-ray. The lower end of the esophagus is seen ending just about the diaphragm which separates the lungs from the abdominal contents including stomach and intestines. The segments seen between the two ends of the esophagus are the vertebral bodies which make up the spinal column. Each vertebral bodies has a pair of ribs arising from it, one to the right and one to the left and they can also be seen. The gap length can either be determined by simple measurement and expressed in centimeters (cms) or the number of vertebral bodies between the two esophageal segments can be counted. In this case the gap length is about five vertebral bodies. If the child weighs about 11 pounds, the distance would be about 5 cm as each vertebral body would be about 1 cm thick. If the child weighed about 15 pounds, however, then the gap length for five vertebral bodies might be about 6 cm. Whichever method of measurement is used, this would be considered an ultra long gap. | Figure 3:  http://www1.umn.edu/eatef/images/fig3.gifhttp://www1.umn.edu/eatef/images/fig4.gifAnother gapogram which shows a seven vertebral body gap length. This was taken from another child with pure EA and, because the upper and lower segments are shorter, the relative gap is somewhat longer. In this case, there is almost no esophagus present in the chest. Truly, an example of a very long gap.  Figure 4:  Another gapogram with even a shorter esophageal segments. In this case, the intrathoracic (chest portion) esophagus is entirely missing. |

A primary repair which joins the two ends of the esophagus together and leaves the stomach below the diaphragm in the abdomen where it belongs, is by far the best solution. A long gap provides the main obstacle to a satisfactory primary repair. Virtually everyone agrees that the child's own esophagus is best and bringing the two ends together is preferable to any substitution using bowel or stomach. Nevertheless, virtually all pediatric surgeons will state that some gaps just are too long and the ends cannot be brought together.

The length of the gap that is too long for a primary repair varies somewhat from surgeon to surgeon. As one of the best known writers in this area, Lewis Spitz, has stated, the difficulty posed by the gap is often in the eye of the beholder ([reference 2](http://www1.umn.edu/eatef/references.html)). If the child is small and the tissues seem more fragile, then relatively short gaps may be judged too long. Certainly, a gap longer than 3 cm has proven to be a significant obstacle to a primary repair. For perhaps 10-20% of infants with some form of EA/TEF, the gap between the two esophageal ends is considered too long to allow them to be brought together. These babies are considered to have a long or very long gap EA and will likely not have a true primary repair.

A note of caution is necessary regarding the gap definitions. We have defined a very long or even ultra long gap as being over 3.5 cm ([reference 3](http://www1.umn.edu/eatef/references.html)). Although there are no other reports of true primary repairs of gaps between 3.5 and 6 cm, one writer has stated that a primary repair is almost always done for them ([reference 4](http://www1.umn.edu/eatef/references.html)). What is meant by a primary repair is that article is that one or more circular myotomies may be done and the stomach may be pulled up part way into the chest. That is why we are so careful about the definition of a true primary repair.

Commonly, a baby with long gap EA will be allowed to grow for several months with the hope that the two ends would also grow together. Although this may occur, typically it does not and the gap grows with the child. Although the relative gap is not any longer because the size of the child has increased, the absolute gap is greater. In some instances, however, the gap does shrink as the two ends grow longer. But setting these fortunate few aside, the problem of the babies with long gap EA remains.