Extrathoracic Lengthening (Kimura Technique)

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Introduction

The treatment of congenital long gap esophageal atresia is extremely challenging. Cases are rare, variable, and oftentimes complex; patients can differ in type of malformation, length of gap, comorbidities, and gestational age. Possible treatment options include delayed primary repair after initial gastrostomy, gastric transposition, partial gastric mobilization, esophageal lengthening or traction procedures such as the Kimura advancement, or the internal or external Foker procedure, colonic or jejunal interposition, esophageal myotomy, and gastric tube techniques [1, 2]. Numerous combinations in timing, chronology, and combination of these methods are possible, and preferences differ between settings, institutions, and surgeons [3].

The so-called Kimura advancement or multistaged extrathoracic esophageal lengthening was initially published by Kimura and Soper in 1992 [4]. It describes advancing a cervical esophagostomy along the anterior thoracic wall subcutaneously in multiple stages until enough length has been gained and primary anastomosis of the native esophagus is achievable (Fig. 6.1a).

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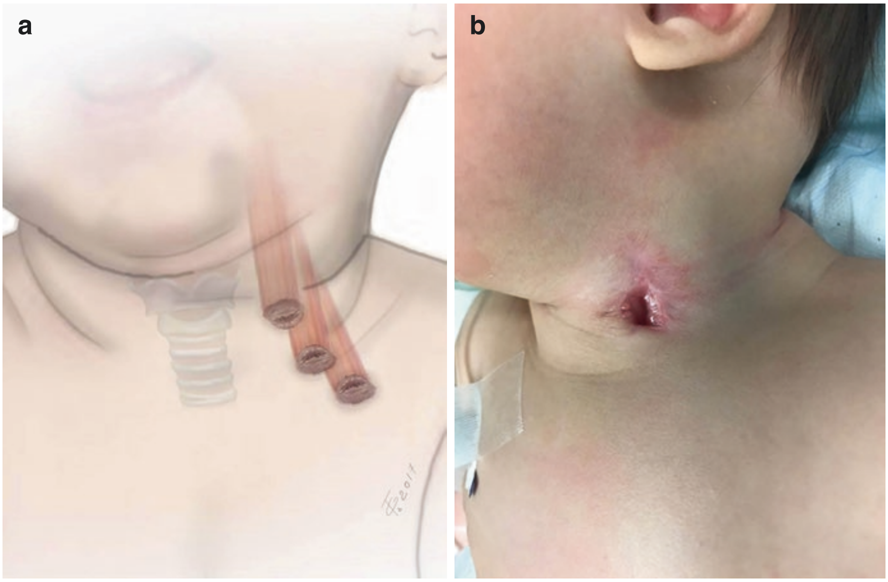

Fig. 6.1 Schematic illustration of the Kimura procedure: staged extrathoracic advancement of the proximal esophagus under traction in a subcutaneous tunnel (**a**). Typical cervical esophagostomy before Kimura advancement is performed (**b**)

Indications: Advantages and Disadvantages

Kimura advancement preserves the upper native esophagus when primary anastomosis is not possible, such as in complex cases of long gap esophageal atresia (LGEA) [4]. Furthermore, the extrathoracic lengthening procedure can be applied in patients who were primarily bridged by creating a cervical spit fistula. The Kimura advancement (KA) is not restricted to neonates and infants but can be performed in patients of all ages [5]. Since the procedure can be adjusted to the patient's specific anatomy, there are hardly any contraindications. However, there are various alternative approaches for management of complex or long gap esophageal atresia. All methods have their specific advantages and disadvantages. Therefore, the patient's parents, caregivers, or legal guardians need to be included in the discussion about what types of techniques are most appropriate in the specific setting, and the potential complications.

Long gap esophageal atresia is a rare condition. Patients present with a variety of characteristics and comorbidities. Overall, treatment strategies either aim to preserve or replace the native esophagus. Unfortunately, high quality and reliable data from prospective, controlled, multicenter research are still not available [1] to date. Even the current treatment recommendations of American Pediatric surgical association (APSA) [1] and ERNICA [6] are mainly based on expert consensus, rather than original scientific data. For KA in particular, there is no data comparing the method to other approaches, therefore evidence is based on small retrospective series and individual case reports. According to American Pediatric Surgical Association (APSA) Outcomes and Evidence-Based Practice Committee [1], 48% of all respondents preferred the upfront use of staged traction repair of the native esophagus such as the Foker technique, while 42% preferred the use of a gastric transposition. In cases of LGEA, delayed primary repair should be considered the best early option for LGEA, followed by esophageal lengthening procedures if unsuccessful [1].

In our experience, Kimura advancement offers advantages in those patients who underwent prior placement of a cervical esophagostomy. In patients born at our institution, we generally try to avoid a cervical esophagostomy and perform an internal traction technique instead until both ends are approximated without tension, ready for a primary anastomosis. However, many of those patients referred to our center have already undergone an unsuccessful attempt at repair and end up with a cervical esophagostomy as a salvage procedure before they are transferred (Fig. 6.1b). The advantage of the cervical esophagostomy is that oral sham feeds can be commenced safely right after the operation, and patients are not dependent on constant Replogle suction of the upper pouch, although modern mobile suction pumps have made even long-term continuous suction of the upper pouch an easy task that can even be performed at home. However, by allowing the patients to take sham feeds orally, the children with a cervical esophagostomy learn to suck and swallow in a timely manner, even before esophageal continuity is achieved. This oral stimulation can be continued throughout the multistage extrathoracic Kimura lengthening process, which may take several weeks to even months. After collecting the milk or formula in the ostomy bag, it can be given via the gastrostomy. In this manner, enzymes in the saliva are physiologically delivered to the gastrointestinal system, possibly providing digestive and immunological benefits. Handling and mobility of the infant are not restricted by Replogle tubes or suction pumps and do not require hospitalization between the procedures. Pending scientific proof, we consider the near physiological feeding situation and absence of a Replogle tube during the stages of Kimura advancement strong advantages of this method, particularly in low-resource settings. The main disadvantages of a cervical esophagostomy are the scar that results from this approach, the sometimes tedious dissection necessary to bring the esophagus back into the chest at the time of anastomosis, the ostomy bag, as well as skin irritation and infection because of the secretions around the esophagostomy.

There are further positive and negative surgical aspects to the extrathoracic esophageal lengthening that the pediatric surgeon should be aware of even though there is a lack of conclusive scientific data for these aspects.

Advantages

With the Kimura advancement, apart from the future intrathoracic anastomosis, all interventions up to that point remain extrathoracic and do not require a thoracotomy or thoracoscopy. For our own cohort, extubation after extrathoracic lengthening surgery is routinely achieved immediately. Also, operative time for the Kimura advancement is much lower (30–60 minutes) compared to an intrathoracic Foker procedure (60–120 minutes).

Disadvantages

From an aesthetic perspective, the multiple stages of Kimura advancement inevitably lead to multiple cutaneous scars in the upper frontal thoracic region. These multiple scars, however, have to be carefully weighed against the scars and postop sequelae other methods, such as an open thoracotomy. In the process of Kimura advancement, we advise for meticulous attention to excise possible remaining mucosa and scarring from the ostomy site to enable the most favorable cosmetic outcome. We also recommend advising the parents in advance of the possible aesthetic implication of multiple surgeries. The effect from the different types and locations of scars in long gap esophageal atresia surgery on patients' psychological well-being has not been evaluated scientifically.

During the Kimura procedure, the proximal esophagus is dissected repeatedly from all surrounding tissue. This extensive preparation along the proximal esophagus may compromise the vascular supply of the upper pouch, which may explain a relatively high leak rate at the time of anastomosis experienced in our own series. However, this effect has not been scientifically evaluated and at this time is purely speculative. A higher leak rate is not only seen with Kimura-type advancement but also in patients who underwent a Foker-type procedure. Impaired vascular supply has in fact been shown for the Foker procedure experimentally in rodents, after short periods of traction of up to 7 days [6–8]. All surgical concepts for the treatment of long gap esophageal atresia involve rather extensive dissection and mobilization of either the proximal, distal, or both stumps, possibly leading to fibrosis and compromised blood supply [9]. While performing the Kimura advancement for esophageal lengthening, repeated suturing of the distal end of the proximal esophagus actually leads to a loss of some length.

Technique

The Kimura advancement surgery is performed in general anesthesia with the patient in supine position with the head extended and turned to the contralateral side of the esophagostomy. The surgical prep area should include chin and mammilla as visual anatomical landmarks for the surgeon. Preoperative endoscopy through the gastrostomy with intraoperative fluoroscopy and a marker at the current upper esophagostomy is recommended to determine the configurations of the upper and lower pouches and to determine the dimensions before commencing the extrathoracic lengthening process. The length of the gap between proximal and distal esophagus is estimated. Continuity of the spit fistula can be demonstrated endoscopically as well. Preoperatively, broad-spectrum antibiotics are administered as a single dose at our center.

The initial elongation after cervical esophagostomy consists of a circular incision surrounding the spit fistula (Fig. 6.2a) und a careful full-thickness dissection esophageal wall from the skin (Fig. 6.2b). Stay sutures are placed in all four quadrants (Fig. 6.2c). The proximal esophagus is dissected circumferentially from the surrounding tissue along its surface and adhesive fibers up to the level of the cricoid cartilage, allowing for elongation of the esophagus [4]. After thorough and careful dissection of the upper pouch, the extent of possible traction is assessed by manually placing the esophagus in traction and determining an appropriate amount of tension (Fig. 6.2d). A new cervical or upper thoracic skin incision is performed at the determined site for the novel esophagostomy (Fig. 6.3a). According to the original description of the method, Kimura et al. recommend to place a Foley catheter in the esophageal lumen at this stage as a useful adjunct for identifying the plane of the esophageal wall [10]. The mobilized esophagus is thus pulled through a bluntly formed subcutaneous tunnel (Fig. 6.3b) to the novel esophagostomy site (Fig. 6.3c) and cutaneous esophagostomy is performed under mild traction using full-thickness sutures (Fig. 6.3d). The previous site of the spit fistula is closed and an ostomy bag

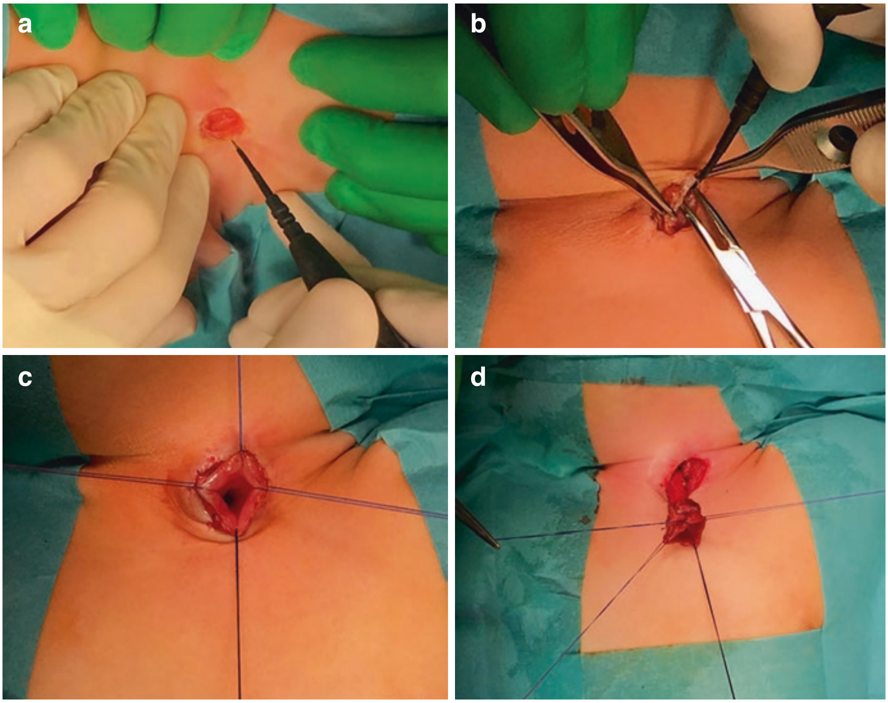

Fig. 6.2 Dissection of the upper esophagus: The mucosa is separated circumferentially from the surrounding skin (**a**) using monopolar electrocautery. Blunt and sharp dissection is used to define the exterior esophageal wall from surrounding soft tissue and adhesions (**b**). Circumferential traction sutures are placed, of which one is marked in a different color or length to avoid rotation of the esophagus (**c**). Dissection of the esophagus up to the level of the cricoid cartilage in order to achieve maximal lengthening (**d**)

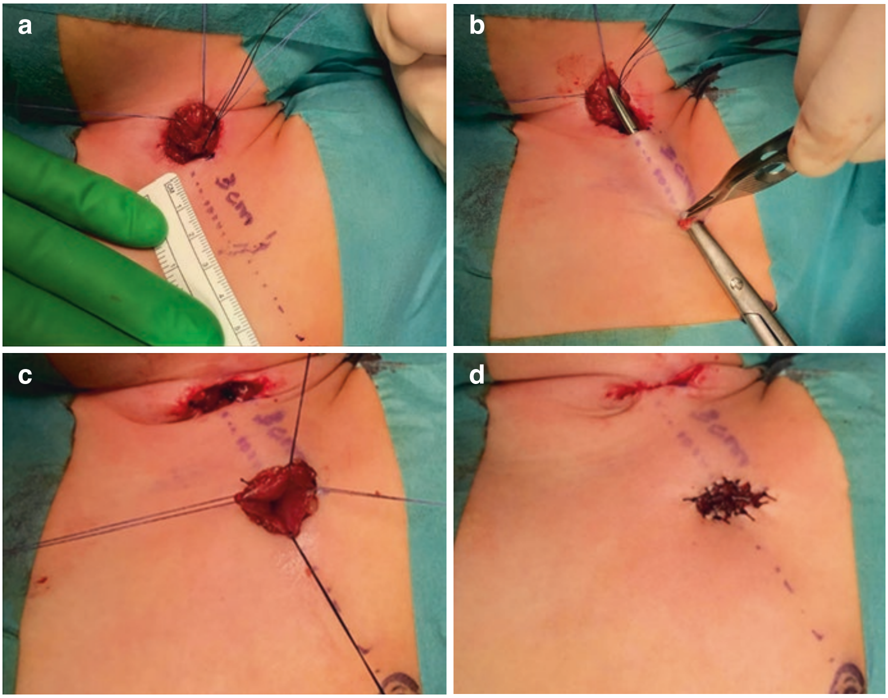

Fig. 6.3 The site of the new esophagostomy is marked (**a**). A skin incision and blunt formation of a subcutaneous tunnel is performed, aiming for approximately the left nipple (**b**). The esophagus is subsequently pulled through the subcutaneous tunnel, carefully avoiding rotation (**c**). Finally, the esophagostomy is performed with interrupted absorbable sutures (**d**)

is placed over the esophagostomy to collect the saliva (Fig. 6.4). Sham-feeds can be commenced on the day of the intervention and collected in an ostomy bag as well. The content of the ostomy bag is then transferred back to the patient via the gastrostomy in regular intervals, so that the content does not spoil. The patient is extubated in the immediate postoperative period and can be transferred to the parents or caregivers when awake. The interval of time between the different stages of surgery is preferably spent at home, unless comorbidities or prematurity contraindicate discharge from inpatient care.

Single *Versus* Multistaged Lengthening

In few cases, primary anastomosis can be achieved after a single extrathoracic lengthening procedure. However, and mostly, several rounds of lengthening are required. In those cases, Kimura advancement is repeated in the same fashion until anastomosis can be achieved. The traction technique on the upper esophagus can easily be combined with other traction procedures, such as an internal or external Foker procedure of the lower pouch. Fluoroscopy is useful in assessing the current

Fig. 6.4 Postoperative care of the lengthened esophagostomy after a Kimura advancement using a colostomy bag to collect the secretions and orally given feeds

gap, placing a marker at the site of the esophagostomy and an endoscope in the lower pouch. Also, it is useful to place a clip as a marker on the tip of the lower esophagus during internal or external traction, so that the approximation of the pouches is visible on conventional chest radiographs. The esophago-esophagostomy after multistaged KA can also be performed thoracoscopically, both from our own experience but also according to other centers [11].

While Kimura et al. advised to wait 2 months to perform the second subcutaneous advancement of the esophagostomy on the anterior chest wall [10], we found that the progression is actually possible as early as 6 days after the first advancement. We generally try to give the esophagus about 2 weeks to recover and lengthen before the next lengthening procedure is performed.

Technical Pitfalls

During the dissection of the upper pouch, it is absolutely vital to avoid perforation of the esophagus. It can be attempted to close an accidental perforation with fine sutures (6-0 monofilament resorbable suture). However, more often than not, the perforation results in a leak and subcutaneous soft tissue infection. In such cases, we would recommend upfront placement of a subcutaneous drain (such as a silicone vessel loop) subcutaneously to avoid fluid collections and abscess formation.

Also, the tension exerted on the tissue should be mild. At this time, there is no objective maximal force that we can recommend, since we currently do not perform intraoperative tension measurements. Therefore, the amount of tension remains a surgeon's subjective choice. In an effort to gain more length, there is a tendency to apply too much force, which can lead to postoperative disruption of the suture line. If this happens, a naso-esophagostomy tube can be placed to maintain the lumen and wait for the disruption to granulate and heal. Ample time should be given in such cases before another lengthening is attempted.

Results and Long-Term Follow-Up

Between 2015 and 2020, our center treated 13 patients with long gap esophageal atresia with a gap of five or more vertebral bodies. In this cohort, a total of 21 Kimura advancement procedures were performed in nine patients, with one to four advancements per patient. The mean number of traction procedures (Kimura and/or Foker procedure) was 2.5 per patient, with a minimum of one Foker procedure ($n = 4$) or one Kimura advancement ($n = 1$) and a maximum of four Kimura advancement plus one Foker procedure ($n = 1$). Of those nine patients that underwent extrathoracic esophageal lengthening of the proximal esophagus, six were simultaneously treated by intrathoracic traction of the distal esophagus. For the seven patients that underwent more than one extrathoracic lengthening of the upper esophagus, the mean interval between Kimura procedures was 11 days (6–13 days) in our center. In our cohort, time between interventions for esophageal traction was considerably shorter than in all other reported case series, in which time between interventions was unanimously around two months as recommended by Kimura et al. [5, 10]. Four out of the five patients were found to have a leak after primary anastomosis following Kimura advancement. However, all of the leaks sealed with time, and without operative intervention.

Of all 12 patients with long gap esophageal atresia treated over the last 5 years, all but one underwent successful native organ esophageal anastomoses with traction procedures (Kimura advancement, Foker procedure, or combinations of both). Only one patient required gastric transposition following a single Kimura advancement due to a change in treatment regimen. In that particular case, no meaningful length of lower esophagus was found. The esophageal stump had been removed and the stump used as a gastrostomy in an outside center.

Mean time between the first traction procedure and the eventual successful esophago-esophagostomy was 42 days (14–92 days) for our patients. All are on full feeds 1–5 years after the last Kimura procedure, except for one, who requires supplementary gastric feeds. Overall, from retrospective analysis of our own cohort treated with extrathoracic lengthening of the upper esophagus, we found Kimura advancement to be feasible and successful as primary treatment option for patients with long gap esophageal atresia, particularly those referred with a cervical esophagostomy.

Discussion

The management of long gap and otherwise complicated esophageal atresia is a challenge that requires a diverse set of strategies. Even though the guiding principle to preserve native esophagus whenever possible is widely agreed upon [3], indications when to depart from this paradigm in the context of long gap esophageal atresia are subject to ongoing discussions. The most appropriate lengthening modality particularly suitable for those patients who previously underwent a cervical esophagostomy is extrathoracic, subcutaneous lengthening of the upper pouch according to the technique described by Kimura.

Literature on isolated extrathoracic lengthening of the proximal esophagus consists mostly of retrospective case series, the largest consisting of 20 patients collected over more than one decade [10–13]. A systematic review of these publications showed that more than 80% of patients treated with Kimura advancement were exclusively on oral feed on last follow-up and less than 50% suffered from reflux [13]. Consequently, the review demonstrates that patient outcome after Kimura advancement is comparable to other treatment strategies for long gap esophageal atresia [1].

Timing of advancement intervals differs among authors. Sroka et al. [5] reported a mean time of 36.3 weeks from first traction procedure to eventual anastomosis for six patients with Kimura advancement and Foker procedures in combination, which is significantly longer than in our experience.

One retrospective two-center study of 15 patients with long gap esophageal atresia treated with traction techniques for the upper and/or lower esophagus found that all patients treated with Kimura advancement eventually underwent a primary esophago-esophagostomy [5]. In this cohort from two European centers, patients treated with the combination of Kimura advancement and Foker procedure suffered from higher infection rates resulting in more thoracotomies [5]. However, patients ($n = 3$) that underwent Foker procedures on both the proximal and distal esophagus rather than extrathoracic lengthening procedures for the proximal esophagus were found to have significantly more severe complications [5]. This corroborates our own findings as described above.

Our approach to the treatment of complicated and long gap esophageal atresia consists in a modular concept tailored to the individual need regarding Kimura advancement as an essential element to achieve esophageal continuity. The high rate of successful esophago-esophagostomy and the rather rapid achievement of primary anastomosis for patients treated with (combined) traction techniques for the upper and lower esophagus together with the dispensability of a Replogle tube make the Kimura advancement preferable to gastric transposition procedures in suitable cases.

One of the major advantages of the Kimura advancement is its technical simplicity and low invasiveness, especially compared to intrathoracic lengthening approaches. It also can be performed in almost any setting, including low-income countries. Furthermore, anesthesia for the cervical Kimura advancement is less challenging, for both patient and staff, compared to that of a Foker procedure. Once cervical esophagostomy and gastrostomy are performed postnatally, the referral to a specialized center can be planned, if necessary. The esophageal anastomosis or further lengthening procedures can be performed electively and adapted to the patient's specific anatomic situation. One of the other main advantages of Kimura advancement is the possibility of sham-feeds, which facilitates a near-physiological feeding experience for the affected child. This way, normal development of the swallowing act is not compromised by extended periods without oral feeds and necessity of Replogle tube. Furthermore, intervals between the surgical stages of the Kimura advancement might be spent at home reducing healthcare costs and improving the families' quality of life.

Conclusion

Multistage esophageal elongation in cases of long gap esophageal atresia using Kimura advancement or a combination of Kimura advancement and Foker procedures is an excellent option to achieve esophageal continuity and preserve the native esophagus when a previous cervical esophagostomy has been performed. Kimura advancement is a valuable pillar in the lengthening procedure providing the possibility of commencements of early feeds and shorter overall hospital stay with relevant advantages for the quality of life for patients and families. Kimura advancement is less invasive and surgically less challenging compared to other approaches and can be performed in almost any setting. From current publications, no single surgical concept in long gap esophageal atresia appears to deliver superior results in terms of long-term outcome variables.

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