



RESEARCH ARTICLE

LONG GAP ESOPHAGEAL ATRESIA WITH OR WITHOUT TRACHEOESOPHAGEAL FISTULA: SEVERAL MANAGEMENT OPTIONS BUT A CONTINUOUS CHALLENGE

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ABSTRACT

Background: The management of long gap esophageal atresia/tracheo-esophageal fistula (EA/TEF) continue to challenge pediatric surgeons and although there were several advances in its management, none of the available options is ideal.

Patient and Methods: This is a retrospective chart review of 42 patients with long gap esophageal atresia treated over a 20 years period. The review included age at diagnosis, sex, type of esophageal atresia, associated anomalies and most importantly the methods of repair and their outcomes.

Results: Out of 42 patients with long gap EA with or without TEF, 23 patients were treated by primary repair of their native esophagus, 15 had esophageal substitution and 4 died before any definitive treatment was performed.

Conclusions: The management of long gap esophageal atresia is challenging and every attempt should be made to preserve the native esophagus. During The second half of the study period we adopted a more conservative approach with the aim of avoiding esophageal replacement using primary repair under severe tension or delayed primary repair after a period of observation. Although this approach is associated with a high incidence of esophageal stricture, we found it to be a better alternative to esophageal replacement in patients with long gap esophageal atresia.

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INTRODUCTION

Repair of long gap esophageal atresia represents a challenge to pediatric surgeons. It must be kept in mind that the baby's own functional esophagus is superior to any esophageal replacement and every attempt should be made to preserve it. To achieve this goal, pediatric surgeons should be familiar with the different techniques to preserve the native esophagus (Al-Shanafey and Harvey, 2008; Paya and Schlaff, 2007; Segulier-Lipszyc *et al.*, 2005; Bagolan *et al.*, 2004; Foker *et al.*, 1997; Boyle and Irwin, 1994). The management of long gap esophageal atresia have changed over the years from an almost standardized esophageal replacement protocol to a more conservative esophageal preservation approach. This study represent our experience with long gap esophageal atresia over a period of 20 years highlighting the different options and the importance of management changes over the years.

Patients and methods

Over a period of 20 years (1993 -2013), 160 cases (91 M: 69 F) of EA/TEF were treated. Forty two (26.25%) of these patients had a long gap EA. Their medical records were reviewed for: age at diagnosis, sex, type of esophageal atresia, associated anomalies, method of repair and outcome. Long gap esophageal atresia was arbitrary defined pre-operatively as a gap length of more than 3cm or 3 vertebral bodies between proximal and distal esophageal pouches or intra-operatively when it was difficult to perform primary esophageal anastomosis or the anastomosis was done under severe tension.

RESULTS

Out of 160 patients with EA±TEF treated during a 20 years period, 42 patients (26.25%) had a long gap EA. There were 29 male and 13 female. Their mean birth weight was 2.45 kg (1100 g – 3800 g). Their mean gestational age was 36 weeks (32-41 weeks). Five cases had a history of maternal polyhydramnios. Three were one of twin and one had a brother with EA.

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The type of the anomaly was as follows: 14 had pure EA, 26 had EA with distal TEF and 2 had EA with proximal and distal TEF. Twenty three patients (54.8%) were treated by primary repair of their native esophagus, 15 (35.7%) had esophageal substitution and 4 (9.5%) died because of sepsis before any definitive treatment was performed.

Patients with long gap pure EA

Fourteen patients had pure EA. Nine patients (9/14) had gastrostomy and cervical esophagostomy as an initial management (Figures 1a and 1b). Three of them died before any definitive repair is performed, and 6 had subsequent esophageal substitution (4 had gastric transposition, 1 had colonic replacement and 1 had gastric tube replacement after initial extrathoracic elongation) (Figures 2a and 2b). The remaining 5 patients (5/14) had gastrostomy only as an initial management. Subsequently, 4 had delayed primary repair and the fifth had colonic replacement. (Table 1)

Patients with long gap EA and distal TEF

Twenty six patients had EA with distal TEF. Three of them (3/26) were managed initially by a cervical esophagostomy and feeding gastrostomy along with fistula ligation, followed after an interval period by colon replacement of the esophagus as a definitive management. Fistula ligation plus a feeding gastrostomy (without a cervical esophagostomy) was the initial management in 12 patients (12/26). Subsequently, 3 had esophageal substitution (2 had gastric tube replacement, 1 had gastric transposition), and 8 had delayed primary repair. The twelfth one died before any definitive repair was performed. The remaining 11 patients (11/26) were managed by primary repair under severe tension. (Table 2)

Patients with long gap EA and proximal and distal TEF

Two patients had EA with proximal and distal TEF: Both had gastrostomy and esophagostomy as an initial management.

Table 1. Management of 14 patients with long gap pure EA

Initial management	No.	Definitive management	No.
Gastrostomy + esophagostomy	9	Gastric transposition	4
		Colonic replacement	1
		Gastric tube replacement (after Kimura extrathoracic elongation and failed delayed primary repair)	1
		Died	3
Gastrostomy only	5	Delayed primary repair	4
		Colonic replacement	1
Total	14		14

Table 2. Management of 26 patients with EA and distal TEF

Initial management	No.	Definitive management	No.
Fistula ligation + gastrostomy + esophagostomy	3	Colonic replacement	3
		Delayed primary repair	8
Fistula ligation + gastrostomy	12	Gastric tube replacement	2
		Gastric transposition	1
		Died	1
Primary repair under severe tension	11	--	
Total	26		15

Table 3. Management of 2 patients with EA and double TEF

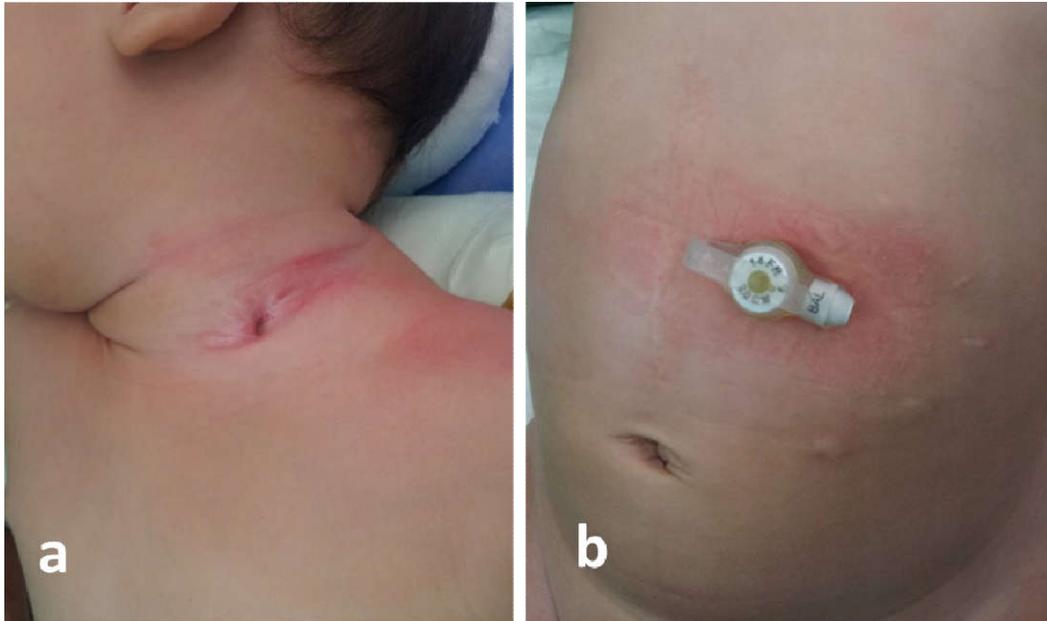
Initial management	No.	Definitive management	No.
Fistula ligation + gastrostomy + esophagostomy + Foker's external traction.	2	Gastric tube replacement	1
		Gastric transposition	1
Total	2		2

Table 4. Management of 15 patients using various esophageal substitutes

Definitive management	No.	Type of the anomaly	No.	Initial management	No.
Gastric transposition	6	Pure EA	4	Gastrostomy + esophagostomy	4
		EA/distal TEF	1	Fistula ligation + gastrostomy	1
		EA/double TEF	1	Fistula ligation + gastrostomy + esophagostomy + Foker's external traction sutures	1
Colon replacement	5	Pure EA	2	Gastrostomy + esophagostomy	1
				Gastrostomy only	1
		EA/distal TEF	3	Fistula ligation + gastrostomy + esophagostomy	3
Gastric tube	4	Pure EA	1	Gastrostomy + esophagostomy + Kimura extrathoracic elongation	1
		EA/distal TEF	2	Fistula ligation + gastrostomy	2
		EA/double TEF	1	Fistula ligation + gastrostomy + esophagostomy + Foker's external traction sutures	1
Total	15		15		15

Table 5. Management of 23 patients using primary repair of the native esophagus

Definitive treatment	No.	Type of the anomaly	No.	Initial management	No.
Primary repair under severe tension.	11	EA/distal TEF	11	---	
Delayed primary repair	12	Pure EA	4	Gastrostomy only	4
		EA/distal TEF	8	Fistula ligation + gastrostomy	8
Total	23		23		12

**Figure 1. Clinical photographs showing cervical esophagostomy (a) and gastrostomy (b)****Figures 2. Clinical photographs showing extra thoracic esophageal elongation**

The two esophageal ends were brought by traction sutures exiting the anterior chest wall and underwent daily traction on these sutures according to Foker's technique. Delayed primary repair was attempted after 3 weeks but failed, and subsequently 1 had gastric tube replacement and the other had gastric transposition. (Table 3)

The method of repair

Esophageal substitution

Fifteen patients were treated by esophageal substitutions; 6 had gastric transposition, 5 had colonic replacement and 4 had gastric tube replacement (Figures 4a, b and c).

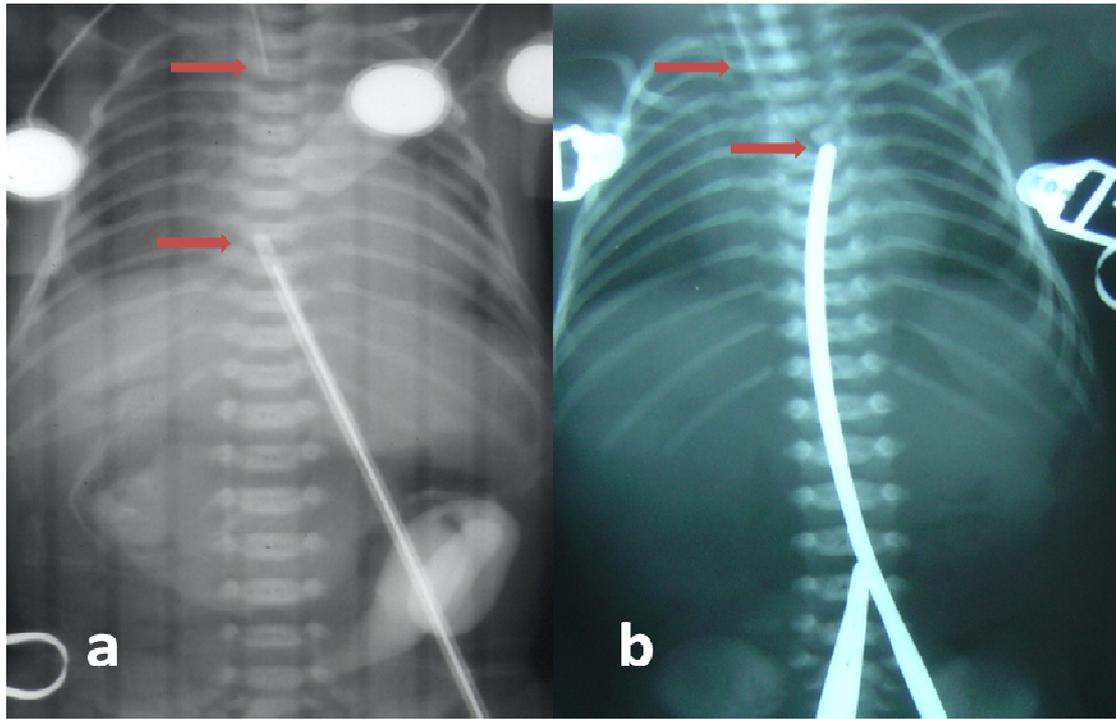
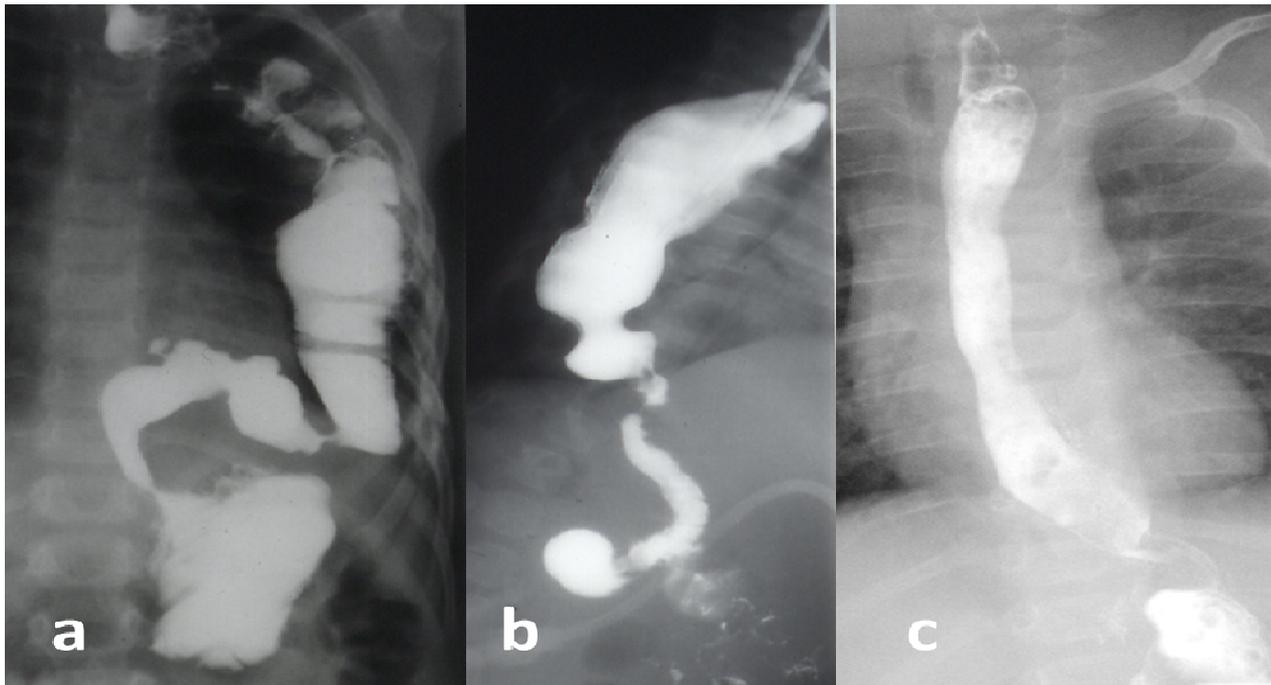


Figure 3. X-rays showing the gap between the upper and lower esophageal pouches (a). Note the decrease in the gap following delay of the repair (b)



Figures 4. Contrast studies showing colonic (a), gastric (b) and gastric tube (c) esophageal replacement

All these patients were treated during the initial 10 years of the study except two of the 4 patients who had gastric tube replacement were treated during the last three years. Fourteen out of these 15 patients were managed initially by a cervical esophagostomy and a feeding gastrostomy which was the management adopted by most institutions for patients with long gap EA during the initial years of our experience.

In one of them delayed primary repair had been attempted after Kimura extrathoracic elongation, but failed and gastric tube replacement was performed. In the 2 patients with EA and double TEF, Foker's external traction sutures were used but didn't add enough length to allow for primary repair and subsequently the 2 patients were managed by esophageal substitution.

Three of those who underwent gastric tube replacement developed leaks at the cervical anastomosis that was treated conservatively and 2 of them developed stricture at the leak site that was treated with dilatation. (Table 4)

Primary repair of the native Esophagus

Out of 23 patients treated by primary repair of the native esophagus, 11 were treated by early primary repair under severe tension. All of them had EA and distal TEF. All developed esophageal strictures of variable severity that were managed successfully by dilatation, except one that required excision of the stricture and end to end anastomosis. Three of these patients developed gastroesophageal reflux (GER) that responded to medical treatment. Delayed primary repair was attempted in 13 patients, only one of them failed and required subsequent gastric tube replacement as a definitive management, leaving 12 successful delayed primary repairs. These included 4 patients with pure esophageal atresia and 9 with EA/distal TEF. All of these patients had feeding gastrostomy and fistula ligation (in cases of EA/distal TEF) as an initial management. None of them had cervical esophagostomy. In 10 patients we used traction sutures to pull the upper and lower esophageal pouches internally towards the chest wall and in 2 patients we used suture approximation of the two esophageal segments without anastomosis. The gap between the two esophageal ends was assessed initially and subsequently at intervals. The decision to make a delayed primary repair was based on the follow-up assessment of the gap and if it was less than 2 cm, delayed primary repair was performed (Figures 3a and 3b).

During the interval period which ranged from 2 to 3 months, all these patients were treated as inpatients with continuous suction of the upper esophageal pouch. Five of these patients developed minor leaks that were treated conservatively and healed. Gastroesophageal reflux developed in 5 of these 12 patients, and was severe enough to require Nissen fundoplication in 3 patients. The other 2 patients responded to medical treatment. Stricture at the anastomotic site was diagnosed in 3 out of these 12 patients and responded to endoscopic dilatation in all of them. (Table 5)

DISCUSSION

Esophageal atresia, with or without associated TEF, is a rare congenital malformation, afflicting approximately 1 in every 3,500 live births (Spitz *et al.*, 1994). In general, the defect can be repaired by primary anastomosis of the proximal and distal esophagus, with concurrent ligation of any TEF, if present. This however is not always possible when the gap between the two esophageal ends is so wide for this anastomosis to be performed. In the past, long gap esophageal atresia was treated by cervical esophagostomy and gastrostomy followed subsequently by esophageal replacement with stomach, colon, or small intestine (Hunter *et al.*, 2009; Tannuri *et al.*, 2008; Spitz, 2006; Spitz *et al.*, 2004; Spitz, 1996; Ure *et al.*, 1995; Ring *et al.*, 1982). Currently and in spite of the recent advances in surgical techniques, long-gap esophageal atresia is still a major surgical challenge.

It is important to be familiar with the different techniques to treat long gap esophageal atresia but every attempt should be made to preserve the native esophagus as the baby's own functional esophagus is superior to any esophageal replacement. There is no consensus available regarding the definition of long gap esophageal atresia. A gap interval between the proximal and distal esophagus measuring greater than 2 cm, or greater than two or three vertebral bodies is the most commonly used definition for long gap esophageal atresia. This however is not accurate as the distance that constitutes long-gap EA may vary according to the skills and expertise of the surgeon performing the repair. Add to this the fact that some of these cases may be anastomosed under tension. Currently, long-gap esophageal atresia is defined as any distance between the esophageal ends in a newborn that is too wide for the surgeon to perform primary anastomosis of the proximal and distal esophagus (Harmon and Coran, 2012).

There are several options to treat long gap esophageal atresia with or without TEF. These include preservation of the native esophagus, or replacement with stomach, colon, or small intestine. During the early years of our experience, we, like others, resorted to gastrostomy and esophagostomy followed by esophageal replacement with stomach, or colon. We did not use the small bowel for replacement. Over the last 10 years, and with the change of trends, we adopted a more conservative approach to preserve the native esophagus. The majority of our patients (23 patients) were treated by this approach. Primary anastomosis of the two esophageal ends during the initial thoracotomy performed in the neonatal period was possible in 11 of our patient, and although the anastomosis was performed under substantial tension, none of them suffered from postoperative leakage. Despite the classic recommendations in the literature regarding the vulnerability of the lower esophagus to dissection, we found that the distal esophagus can be mobilized down to the esophageal hiatus without any effect on healing. Similar findings have been reported by other surgeons (Harmon and Coran, 2012). Anastomotic stricture developed in all these patients, and gastroesophageal reflux developed in three of them. However, the majority of these problems were successfully treated non-surgically and did not negatively affect the final outcome. All these patients were put postoperatively in a head-flexion position and were kept paralyzed under mechanical ventilation for a period of 5 days. This postoperative management protocol was adopted to decrease the traction force on the anastomosis which is already under tension. Livaditis *et al.* (1972) suggested the use of myotomies to provide extra length of the native esophagus. None of our patients had myotomies and we like others feel that myotomy may have devastating effects including the development of esophageal diverticulum and should be avoided (Tannuri *et al.*, 2003).

Spontaneous growth of the esophagus continue to occur during the first months of life, possibly induced by reflux of gastric contents into the lower pouch (Foker *et al.*, 1997). This has been the basis for delayed primary repair of the native esophagus after a period of observation when primary repair is not possible at the initial operation. Many techniques have been described to shorten the gap between the two esophageal ends and facilitates subsequent primary anastomosis. Traction

on the two esophageal ends have been shown to act as a growth stimulus in itself and also to prevent adhesions from hindering the growth of the esophagus. Foker's external traction was used in two of our patients and Kimura's extra thoracic lengthening was used in one (Sroka *et al.*, 2013; Kimura *et al.*, 2001; Foker *et al.*, 1997). In all three patients, these techniques did not add enough length to allow for a primary repair, and the patients developed esophageal leak and ultimately required esophageal replacement. These results can be attributed to our limited experience with these techniques rather than to the usefulness of the techniques themselves. In 10 of our patients we used internal traction sutures to pull the upper and lower esophageal pouches internally towards the chest wall and in 2 patients we used suture approximation of the two esophageal segments without anastomosis. These 12 patients subsequently underwent successful delayed primary repair.

Many authors have reported high incidence of feeding troubles after delayed primary repair (Friedmacher and Puri, 2012; Cavallaro *et al.*, 1992). A meta-analysis of 451 newborns with long gap esophageal atresia managed with delayed primary repair concluded that this method of repair provides good long-term functional results but with high incidence of gastroesophageal reflux and anastomotic strictures (Friedmacher and Puri, 2012). The findings in our study came in accordance with these findings as 8 of the 12 patients treated by delayed primary repair suffered from these complications. However, they were recognized and managed early with no long term negative consequences.

Patients with long gap esophageal atresia managed initially by gastrostomy and no esophagostomy require careful attention to the upper pouch while awaiting definitive repair. Although in selected cases the patient can be discharged home with a repleg tube in situ for suctioning, we were hesitant to adopt such a protocol and kept all patients in hospital taking in consideration the risk of aspiration pneumonia (Aziz *et al.*, 2003; Hollands *et al.*, 2000). This approach was costly but safe.

In our series, the number of patients who had colonic, gastric or gastric tube esophageal replacements were small to compare between them. Each of these methods of replacement has its associated limitations and potential complications that may restrict its suitability for a particular group of patients. A number of complications have been reported in association with esophageal replacement, including anastomotic leak, stricture, reflux, diarrhea, and colonic redundancy. Add to this, the possibility of severe complications such as graft necrosis, colocolic anastomosis leak, and delayed gastric emptying (Lee *et al.*, 2014; Tannuri *et al.*, 2007; Ludman and Spitz, 2003; Anderson, *et al.*, 1992). In our small number of colonic esophageal replacements, it was associated with redundancy and dysphagia on long term follow-up. Gastric transposition and in spite of the relatively large size of the stomach initially, did not cause long term complication, and on follow-up, the stomach became like a tube with no respiratory or feeding difficulties (Hunter *et al.*, 2009; Spitz *et al.*, 2004). Gastric tube esophageal replacement was associated with a high incidence of cervical leak and although these leaks healed

conservatively, they were followed by a stricture at the anastomosis site (Tannuri *et al.*, 2008).

We acknowledge that our study numbers are limited and that we cannot make firm conclusions regarding the choice of repair. However, based on our institutional experience, we feel that delayed primary repair or primary repair under tension may be the preferred initial method of reconstruction in long-gap EA. Gastric transposition is the alternative for patients in whom primary repair with native esophagus is not technically feasible.

Conclusion

In conclusions, the treatment of long gap esophageal atresia remains a major surgical challenge. Although several surgical options are available for the management of this anomaly, controversy exists regarding the optimal approach. The use of native esophagus in the form of primary repair under severe tension or delayed primary repair, although associated with a high rate of stricture and GER, is the preferred approach. Although this study is limited by numbers, gastric transposition may be favored for patients in whom primary repair with native esophagus is not technically feasible.

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