

A07/S(HSS)a

**2013/14 NHS STANDARD CONTRACT
ENCAPSULATING PERITONEAL SCLEROSIS TREATMENT SERVICE (ADULT)**

PARTICULARS, SCHEDULE 2 – THE SERVICES, A – SERVICE SPECIFICATION

Service Specification No.	A07/S(HSS)a
Service	Encapsulating peritoneal sclerosis treatment service (Adult)
Commissioner Lead	
Provider Lead	
Period	12 months
Date of Review	

1. Population Needs

1.1 National/local context and evidence base

The exact incidence and prevalence of encapsulating peritoneal sclerosis (EPS) is not clearly defined. Reported occurrence rates in countries vary widely. Rigby and Hawley reported EPS in 54 of 7,374 patients (0.7%) in Australia (Rigby et al. 1998). In 1985, a report covering 19 European countries, described an EPS prevalence ranging from 0.3per 1000 patients in Spain to 3.1 per 1000 patients in Belgium (Kramer et al. 1985). The most recent overall EPS incidence in Japan is reported to be 2.5%, but is proportional to duration of dialysis; in those on peritoneal dialysis (PD) for less than 5 years it is 0.7%, rising to 5.9% after 10 years (Kawanishi et al. 2005).

In 2006, there were approximately 19,378 individuals on dialysis in the UK of which 15,511 were on Haemodialysis (HD) and 3,867 were on Peritoneal Dialysis (PD) (Renal Registry data 2008). It is estimated that 3.3% of the patients on PD will develop EPS and so approximately 150 cases annually could be expected, taking into account a 20-25% recurrence rate.

There could, however, be a wide variability in the caseload depending on several factors which include variation between units, diagnostic differences including non-diagnosis, reduction in uptake of PD as a modality, better surveillance, earlier discontinuation of PD and increased transplant rates. It is anticipated that ten years from now EPS will no longer be the problem that it is currently. An estimate for workload over the next 10 years would be a starting caseload of 50 per year, increasing to 100 to a maximum of 150 new cases at five years and falling off thereafter to 30 to 50 cases in 10 years.

Evidence for the medical treatment of EPS, as opposed to surgical treatment, is still unclear. The majority of literature is based on anecdotal case reports with a paucity of clinical trial data. The best data come from Japan, where kidney transplantation is uncommon so patients spend many years on PD, and are thus more likely to develop the disease. One study from Japan recommends treatment guidelines based on a retrospective analysis of 256 patients with EPS (Nakamoto et al. 2002). This report advocated initial peritoneal rest with subsequent steroid treatment and surgical intervention in the obstructive stage of the disease. Following this treatment regime, the two-year survival rate was 73% whereas it was only 48% in those not treated. Another Japanese review article (Kawaguchi et al. 2005) reviewed the summary of alternatives for treatment for EPS and gave surgery the highest level of evidence/importance in the management of EPS.

2. Scope

2.1 Aims and objectives of service

The Encapsulating Peritoneal Sclerosis Surgical Service (EPS SS) provides surgical treatment for encapsulating peritoneal sclerosis (EPS). EPS, also referred to as sclerosing peritonitis, is a complication arising from long term use of peritoneal dialysis (PD), generally over five years, and recurrent episodes of bacterial peritonitis. EPS is being increasingly recognised in the PD group, as waiting times for transplants increase and duration of PD increases.

The aim of the EPS SS is to provide a centralised service for the surgical management of this group of patients. EPS is a condition associated with significant morbidity and mortality and with poor outcomes if not recognised early and treated. As it is a rare condition, experience with management in the UK has hitherto been limited leading to poor outcomes and a high mortality rate. Excellent outcomes with surgery have been shown in Japan. With centralising treatment in specified national centres, experience will be consolidated leading to better patient outcomes, mirroring the Japanese experience.

All patients with EPS who are fit for major surgery will be eligible:

Objectives:

- to provide an exemplary and comprehensive service for all eligible referred patients with EPS
- to provide expert diagnosis of EPS utilising the most up-to-date validated diagnostic tools and knowledge
- to enable the expert management of patients with confirmed EPS through the use of the most up-to-date clinical protocols for prescribing and symptom management
- to offer clinically appropriate consideration and provision of surgery within the EPS patient pathway

- to effectively monitor patients to ensure optimal functioning for the patient with regards to their EPS
- to operate a rolling programme of clinical audit to test current best practice and inform the evolution of care in EPS
- to provide care with a patient and family centred focus to maximise the patient's experience of care within the nationally designated providers
- to be recognised as the leading clinical services and a source of expert advice for the diagnosis and management of EPS within the NHS
- to support local healthcare providers to manage patients with EPS whenever it is clinically appropriate and safe to do so
- to provide high quality information for patients, families and carers in appropriate and accessible formats and mediums
- to develop the experience, knowledge and skills of the multi-disciplinary team (MDT) to ensure high quality sustainable provision.

The strategic objectives of the service are to provide a national referral centre from two sites; Central Manchester University Hospitals NHS Foundation Trust (CMUH) and Cambridge University Hospitals NHS Foundation (CUH) for the surgical management of the condition, consolidating experience and improving outcomes whilst at the same time, evaluating the scale of the problem nationally in the population at risk. The exact incidence of this condition in the PD population in the UK is still unascertained. It is hoped that by centralising treatment, all patients will become known, and over a period of time the exact incidence will be determined. The purpose of the service is to provide the correct and definitive treatment of EPS in all patients diagnosed with the condition. The goal is to return patients to their home environment, with a good quality of life - eating, gaining weight and symptom-free.

2.2 Service description/care pathway

The initial patient referral is assessed against a set of treatment criteria by the consultant renal surgeon. If the case is straight forward the referral is accepted and the patient admitted as an inpatient to the ward. If the case is more complicated the patient may be transferred for an assessment of their case by the consultant renal surgeon, consultant nephrologists and consultant intensivists with input from the dietician and pharmacist. If the patient is assessed as being suitable for surgical intervention, the patient is admitted. If the patient is unsuitable for surgical intervention, the patient will be returned to their referring hospital either with a treatment plan or on a palliative care pathway.

The service is delivered on an inpatient basis. Typically patients will stay for eight weeks; however this can be several months. The preoperative process consists of the patient commencing haemodialysis, total parenteral nutrition (TPN), and being prepared for surgery.

The initial surgery usually consists of a decortication (often termed peritonectomy, although the peritoneum is seldom removed) and enterolysis. After the surgery, the patient may need to return to theatre for a further laparotomy. Once the patient has been discharged the patient may suffer a recurrence of the disease and be

readmitted for further surgery.

Some of the patients have a stoma formed as part of the surgical procedure. In order for the bowel to heal the patient will be left with a stoma for 6 months or longer. The patient will then be readmitted for a stoma reversal procedure.

Patients can return from theatre with an open wound. Vacuum-assisted therapy (VAC) therapy is commenced. A foam dressing is cut to shape and placed in the wound. A tube is attached to the foam which incorporates a suction device. Attached to the other end of the tube is a canister. The wound area is then sealed with a sticky film.

The aim of the therapy is to accelerate healing by increasing oxygen flow to the wound. The area should remain clean of harmful bacteria and any other substances due to the film. Preventing these harmful bacteria from entering the wound should decrease healing time and increase the production of chemicals that encourage tissue growth. These dressings are changed every 48 hours.

The aim of the service is to surgically treat patients with a confirmed diagnosis of EPS. Success will be defined as patients who:

- no longer require TPN feeding,
- have recovered from malnutrition and can return to a normal diet
- be treated for their on-going medical conditions as an outpatient rather than an inpatient.

The service will be staffed with a range of suitably qualified health professionals which will include the following people:

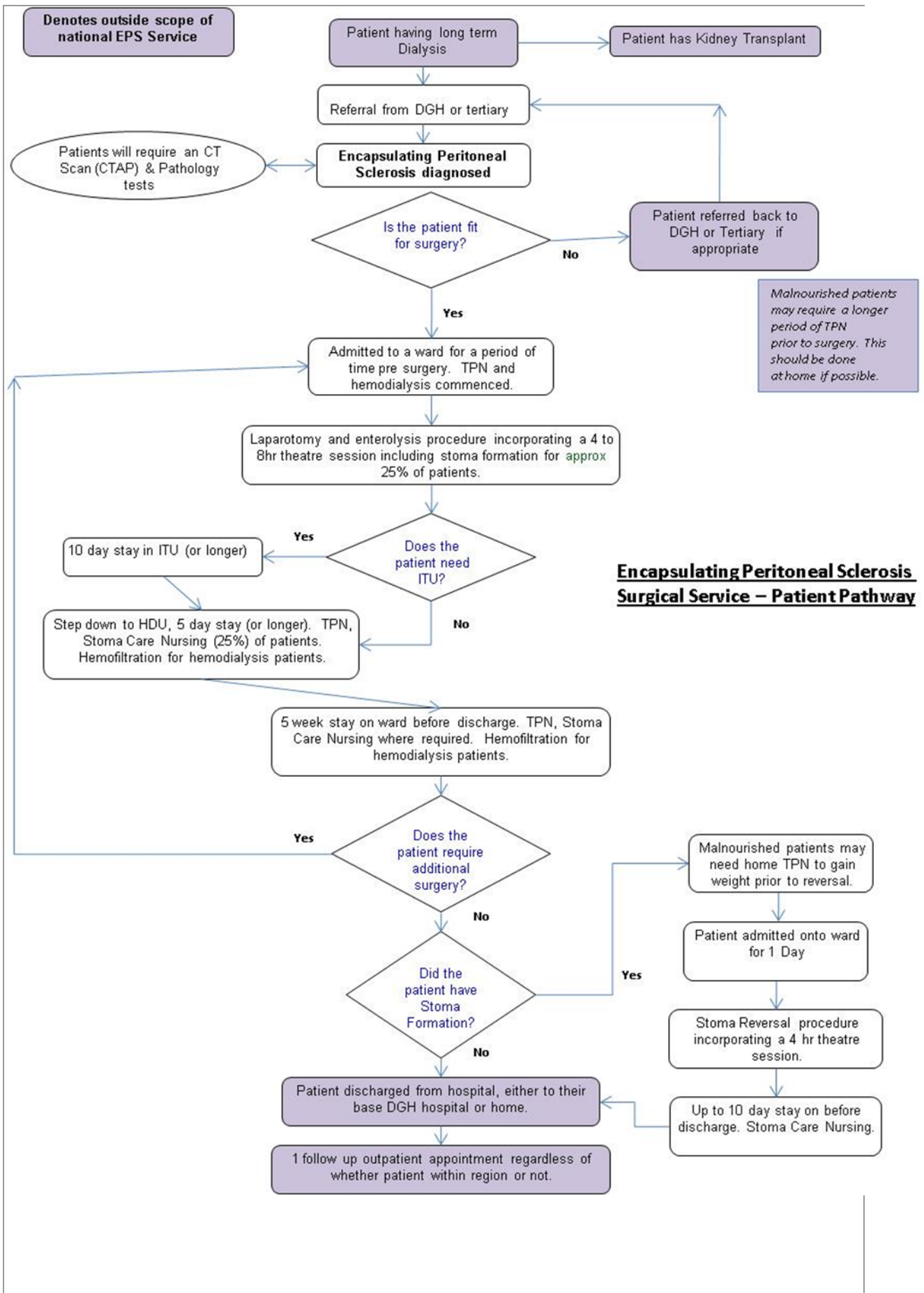
- a consultant surgeon and members of their team
- a consultant nephrologist and members of their team
- dialysis nurses
- ward nurses that can provide levels of care equivalent to those that patients on an intensive therapy unit (ITU) receive
- specialist dietetics input
- specialist pharmacist input
- specialist VAC therapy nursing input
- occupational therapy

Patients' views are sought via trust patient engagement schemes. Care delivered by the EPS service providers must be of a nature and quality to meet the care standards, specification and agreement for the service. It is the trust's responsibility to notify the commissioner on an exceptional basis should there be any breaches of the care standards. Where there are breaches, any consequences will be deemed as being the trust's responsibility.

Patients must be managed in line with the specification and care standards. Any deviation from these which has not been approved by the NHS England is at the trust's risk both clinically and financially. It is the trust's responsibility to inform the commissioners of any such non-approved deviations on an exceptional basis.

Where a patient's presentation challenges the assumptions that underpin the specification, service standards and contractual arrangements it is the trust's responsibility to inform the commissioners on an exceptional basis, prior to any treatment (except for emergency treatment) so that the implications of the patient's requirements can be considered. This does not affect situations where the Individual Funding Application process applies.

Adopted



2.3 Population covered

This service covers patients resident in England, patients resident in the European Union or otherwise eligible for treatment in the NHS under reciprocal arrangements.

Patients resident in Wales, Northern Ireland and Scotland (from 2011-12) are not part of this commissioned service and the trust must have separate arrangements in place for these patients.

International patients outside of the EU (except where reciprocal arrangements have been made by the UK government) are not part of the nationally commissioned service and the trust must have separate arrangements in place for those patients.

2.4 Any acceptance and exclusion criteria

The service is accessible to all patients with EPS regardless of sex, race, or gender.

Providers require staff to attend mandatory training on equality and diversity and the facilities provided offer appropriate disabled access for patients, family and carers.

When required the providers will use translators and printed information is available in multiple languages.

The provider has a duty to co-operate with the commissioner in undertaking Equality Impact Assessments as a requirement of race, gender, sexual orientation, religion and disability equality legislation.

The hospital must be compliant with the Department of Health rules around provision of single sex accommodation for patients. The hospital must have a robust policy for Equality and Diversity and regularly undertake Equality Impact Assessments.

Referral criteria, sources and routes

All patients suspected of having EPS based on a history of a long period of peritoneal dialysis, usually more than five years (sometimes much less) with recurrent attacks of peritonitis with chronic abdominal symptoms of pain, sub acute obstruction along with low albumin, raised C-Reactive Protein (CRP) and refractory anaemia.

Standard blood tests and an abdominal computerised tomography (CT) scan should have been completed before the referral is accepted. A referral will be accepted from any referring consultant, primarily a renal consultant, but also from any other speciality which has entertained a diagnosis of EPS.

The referral will be handled by the EPS coordinator or duty surgeon. An initial decision will be made on the acuteness of the situation. If emergency surgery is required, an urgent transfer will be arranged after input with the intensive care team.

Exclusion criteria

All patients will be considered, except for patients with significant co-morbidity or with a very high risk of mortality. By and large all patients will be transferred for a multi-disciplinary evaluation unless the clinical situation is too grave for transfer.

Response time & detail and prioritisation

No waiting times envisaged, with immediate response to referrals. Patients will be prioritised according to clinical needs.

2.5 Interdependencies with other services

The key stakeholders of this service are any other trust that refers a patient. The service accepts tertiary referrals and will ensure that the patients referring consultant and GP are communicated with on a regular basis.

The service relies upon good working relationships between the 2 national providers at:

- Cambridge University Hospital NHS Foundation Trust;
- Central Manchester & Manchester Children's University Hospitals NHS Trust.

The EPS service is interdependent on the following services:

- nephrology service
- transplant service
- referring consultants/hospitals
- general practice
- dietetics service
- occupational therapy services
- pharmacy services
- interpreter services
- specialist nurse services
- transport services (transfer between hospitals).

There are no relevant networks and screening programmes for this service.

3. Applicable Service Standards

3.1 Applicable national standards e.g. NICE, Royal College

The nationally designated EPS providers must be fully integrated into their trust's corporate and clinical governance arrangements.

The commissioners and service will conduct a formal service review at least every

six months. These take place as a combined review with other nationally designated services.

The service will operate with regard to the agreed service standards.

4. Key Service Outcomes

- Mean survival post surgery
- % patients no longer requiring parenteral nutrition

5. Location of Provider Premises

The service will be delivered from two centres in England:
University Hospitals Cambridge NHS trust
Central Manchester NHS Foundation trust.

Appendix 1 – Care Pathway

THE SURGICAL PATHWAY FOR EPS SURGICAL SERVICE (EPS-SS)

THE PRINCIPLES OF SURGERY

Surgical management in EPS endeavours to tackle the underlying patho-physiological process i.e. the thickened, sclerotic membrane that encapsulates the gut (often described as a cocoon). With encapsulation, the gut is shortened and peristalsis is restricted leading to mal-absorption. Sub-acute and recurrent obstruction occurs and in advanced cases, there is complete mechanical obstruction, internal herniation, and strangulation leading to peritonitis, septic shock and death.

The thickened membrane is the result of a chronic inflammatory process with associated persistent elevation of inflammatory markers, and anaemia. Haemorrhage may occur from this inflamed thickened membrane leading to an acute abdomen and the need for regular blood transfusions.

With on-going EPS, the patient is hospitalised for long periods of time and requires total parenteral nutrition (TPN). Episodes of sepsis, such as aspiration pneumonia (a consequence of intestinal obstruction), may require intensive management on an intensive care unit. Thus, the cost of supporting patients is significant, and may be dramatically reduced with appropriate intervention.

Surgery should be properly timed so that it is considered after an adequate trial of medical therapy and before the patient is too decompensated. It should be performed in a setting where the surgical team has experience of treating EPS, understands the condition and has proper peri-operative renal, transplant and

intensive care support.

INDICATIONS FOR SURGERY

A diagnosis of EPS, based on:

- history of PD (the longer the period on PD, the higher the index of suspicion)
- clinical symptoms of EPS
- CT confirmation of the diagnosis
- lack of response to medical treatment.

Medical treatment involves converting the patient from PD to haemodialysis, and treatment with steroids and tamoxifen together with TPN if there are features of obstruction. Failure of medical treatment, and indications for surgery, are indicated by the following:

- symptomatic (vomiting, pain), raised CRP, persistently low albumin, lack of weight gain
- bowel obstruction (acute and recurrent sub-acute)
- intra-peritoneal haemorrhage
- peritonitis.

PREOPERATIVE PREPARATION

Patients are admitted 10 to 14 days prior to surgery for preoperative evaluation and nutritional support. A Hickmann line is inserted for TPN and parenteral nutrition is given for at least 10 days prior to surgery and continued into the postoperative period. Patients with EPS are malnourished and preoperative nutrition is vital. Other requirements include blood transfusion, if anaemic, haemodialysis (often daily if on TPN), and chest physiotherapy.

Most patients will require computerised tomography (CT) scans to assess the peritoneal cavity, in addition to chest X-ray, echocardiogram (ECG), blood tests, liver function tests and a coagulation profile.

SURGERY

Surgery is a difficult, painstaking procedure that may take four to eight hours. For optimum outcome, surgery should be done electively or semi-electively on a planned list. Surgery carried out as an emergency is associated with a high mortality and morbidity from enteric perforations and haemorrhage.

ANAESTHESIA

Sick patients with EPS are difficult anaesthetic candidates and need an experienced anaesthetist capable of dealing with renal failure patients, as there may be the need for significant transfusion of blood and blood products and fluids intraoperatively. Broad spectrum antibiotics and anti-fungal drugs are given at induction of anaesthesia, and care is taken to avoid aspiration.

THE OPERATION

Careful laparotomy, drainage of ascitic fluid (which is sent for culture cytology) and careful dissection of inflamed fibrotic tissue from the abdominal wall (the resected material also been sent for histology, culture and special tests). The thickened inflamed tissue is also dissected from bowel loops. The aim is to remove all thickened and diseased sclerotic membrane from the bowel wall, but a balance has to be achieved between stripping off the encapsulating membrane and avoiding inadvertent perforation of the bowel. Dissection is continued until the entire small bowel is freed right from the duodeno-jejunal flexure (Ligament of Treitz) to the ileo-caecal junction. The thickened membrane from the pelvis, over the rectum, the colon and the liver and the under surface of the diaphragm is removed if it comes away without difficulty or bleeding. If gastric fullness is a major symptom and there is thickened membrane over the stomach, at least some of it should be removed to free the stomach.

If there is an inadvertent enterotomy, a proximal diverting stoma is necessary since a simple closure, or resection and closure, will invariably break down. In an emergency situation, with bowel gangrene or with significant intra-abdominal sepsis, a laparostomy may be used with delayed abdominal closure. In this situation, the patient is left ventilated, and the abdomen washed out every 48hrs and closed once clean.

POSTOPERATIVE MEDICAL MANAGEMENT

An intensive care bed is mandatory in most patients. Patients may need to be ventilated and haemofiltered/haemodialysed after surgery mainly for hyperkalemia and to make room for TPN. Parenteral nutrition is continued until bowel movement returns and the patient is able to take orally. Nutritional supplements and nasogastric feeding may need to be continued till albumin levels improve. Further transfusions/iron supplements/erythropoietin is continued.

Antibiotics and anti-fungal therapy is continued for five days, and is guided by cultures taken in theatre in during the post-operative period.

In the early post-operative period any intra-abdominal collections are drained and bleeding may require re-operation. Small proportions of patients rapidly develop recurrent EPS and require repeat assessment and surgery.

Tamoxifen is thought to be of benefit in preventing recurrence and is started when the patient is able to take food orally and continued for up to two years. Systemic steroids (prednisolone) may also be given for 6 months.

RECURRENT EPS

Recurrence occurs in about 25%(Kawanishi et al. 2006), and a small proportion of patients will experience multiple episodes of recurrence. The principles of surgical management remain the same as in the primary case, and early surgery is appropriate.

Adopted