Clinical Commissioning Urgent Policy Statement
Surgical treatment of complex primary cardiac tumours in children [170123P]

Commissioning Position

Summary

Surgical treatment is recommended to be available as a treatment option through routine commissioning for complex primary cardiac tumours in children within the criteria set out in this document.

Information about surgical treatment for complex primary cardiac tumours in children

The intervention

Currently in England, surgical resection of most cardiac tumours takes place in designated Children's Cardiac Surgery centres. Rarely, resection of very large tumours may require the backup of mechanical cardiac support or of transplantation. Some patients may be referred to one of the two paediatric cardiothoracic transplantation centres for assessment and may be listed for transplantation.

Committee discussion

The condition

The majority of primary cardiac tumours are benign rather than malignant (cancerous). By nature of their outright size and/or their precise location and relationships to adjacent structures, cardiac tumours may alter the structure and function of surrounding cardiac tissue. This can cause: abnormal cardiac rhythms, problems with blood flow through the heart (either obstruction of inflow or outflow from the heart) and malfunction of the cardiac valves. There may be a risk of embolic phenomena or sudden cardiac death due to the tumour.

Current treatments

Transplantation can be lifesaving but carries risks, including the risk of operation and the risks of lifetime immune suppression, including an increased risk of malignancy. Hence resection without transplant is the preferred option. However, median survival after paediatric heart transplant is about 20 years, with good quality of life.

Comparators

For patients deemed to need tumour resection as set out in the pathway below, the comparators to tumour resection are ongoing medical management including palliative care pathway or proceed directly to transplant.

Clinical trial evidence

NHS England has considered the evidence submitted as part of the preliminary policy proposal to establish the urgent clinical commissioning policy statement, including the clinical criteria for initiating the intervention. This includes up to three of the most clinically impactful publications, identified using a literature search strategy defined by the clinical lead. These publications are summarised below.

Publication 1 Walter et al (2016)

Walter et al (2016) reported a series of 47 consecutive patients with primary cardiac tumours treated at a single institution in Germany between 1986 and 2012. The tumours included rhabdomyoma (13 patients), fibroma (12 patients), teratoma (9 patients) and myxoma (8 patients) as well as other rarer types. The patients were aged from one day to 17 years. The indications for operation were stated as hemodynamic/respiratory compromise, severe arrhythmia, and a significant embolization risk. A 5 month old infant required ventricular assist device followed by heart transplant at 17 days post operation. Two patients died shortly after the operation, one aged 5 months and the other aged 8 months at operation; there was one late death in a 16 year old patient who died of lymphoma seven months after operation. At a mean of 11 years' follow up, all survivors were well, free of tumour-related symptoms and tumour progression and recurrence, even when tumour resection was incomplete. It is not clear whether this centre evaluated other patients with tumours who were not offered surgery.

Publication 2 Nathan et al (2014)

Nathan et al (2014) reported a series of 20 consecutive patients with ventricular fibroma treated at a single institution in the USA between 1990 and 2013. The patients were aged from 5 months to 12 years. Eighteen of the patients had presented with arrhythmia, one with outflow obstruction and one was picked up during routine evaluation. All tumours were resected successfully with no recurrence of arrhythmia; follow up ranged from one month to 14 years. No patient died. It is not clear whether this centre evaluated other patients with tumours who were not offered surgery.

Publication 3 Padalino et al (2014)

Padalino et al (2014) reported a series of 52 children with cardiac tumours treated at a single institution in Italy between 1982 and 2009. The diagnosis was prenatal in 35% of patients. The tumours included rhabdomyoma (32 patients, of whom 22 had tuberous sclerosis complex), fibroma (8 patients) and myxoma (3 patients). Forty-one patients were managed medically and 11 underwent surgical resection. At a mean follow up of seven years, two patients had died of complications from heart transplant; the others were alive and well.

Adverse events

As noted above the major adverse events, in addition to the standard risks of open heart surgery involving resection of heart muscle, are peri operative death and heart failure requiring assist devices and / or transplant.

Implementation

Criteria

This policy covers clinical instances where the presence of the tumour alters the structure and function of the surrounding cardiac tissue either through outright size or by directly impacting upon vital, adjacent structures. The decision to operate is a matter of clinical judgement, balancing benefit and risk. The criteria to consider at multidisciplinary assessment include the following:

- abnormal cardiac rhythms not controlled by medical therapy;
- problems with blood flow through the heart (either obstruction of inflow or outflow from the heart) severe enough to compromise cardiac output;
- malfunction of the cardiac valves with a risk of embolic phenomena;
- risk of sudden cardiac death due to the tumour.

Surgery should be considered only where the balance of risks and benefits (informed by expert paediatric cardiology input to the multidisciplinary team (MDT)) favour surgery over optimum medical management.

The proposed patient pathway is as follows (see appendix):

1. Patient is identified with a cardiac tumour.
2. Referral to local paediatric cardiac surgery service.
3. Investigations and MDT discussion at local paediatric cardiac surgery service.
4. MDT outcomes:
    a. Tumour does not need resection – continue non-surgical management
    b. Needs resection and suitable for resection at local paediatric cardiac centre – proceed to operation
    c. Unsure, or needs resection but not suitable for local paediatric cardiac centre – refer to another UK paediatric cardiac surgical centre, which may (see para 6) or may not (see para 5) be a transplanting centre.
5. If patient referred to a non-transplanting paediatric cardiac surgery centre: discussed at MDT with outcomes as follows:
    a. Tumour does not need resection – continue non-surgical management
    b. Suitable for resection – proceed to operation
    c. Resection not feasible – refer to UK transplanting centre: see para 6
6. Assessment at transplant centre; transplant MDT outcome:
    a. Tumour resectable without need for heart transplant
    b. Tumour resectable but only with subsequent (or simultaneous) heart transplant – discuss with other UK transplant centre to confirm opinion before listing (advice of international experts may be sought at this point).
    c. Tumour deemed not resectable and patient not eligible for transplant – discuss with other UK transplant centre.
        i. If both centres agree not for surgery or transplant, patient is referred back to the local paediatric cardiac surgical centre for medical management or supportive care pathway.

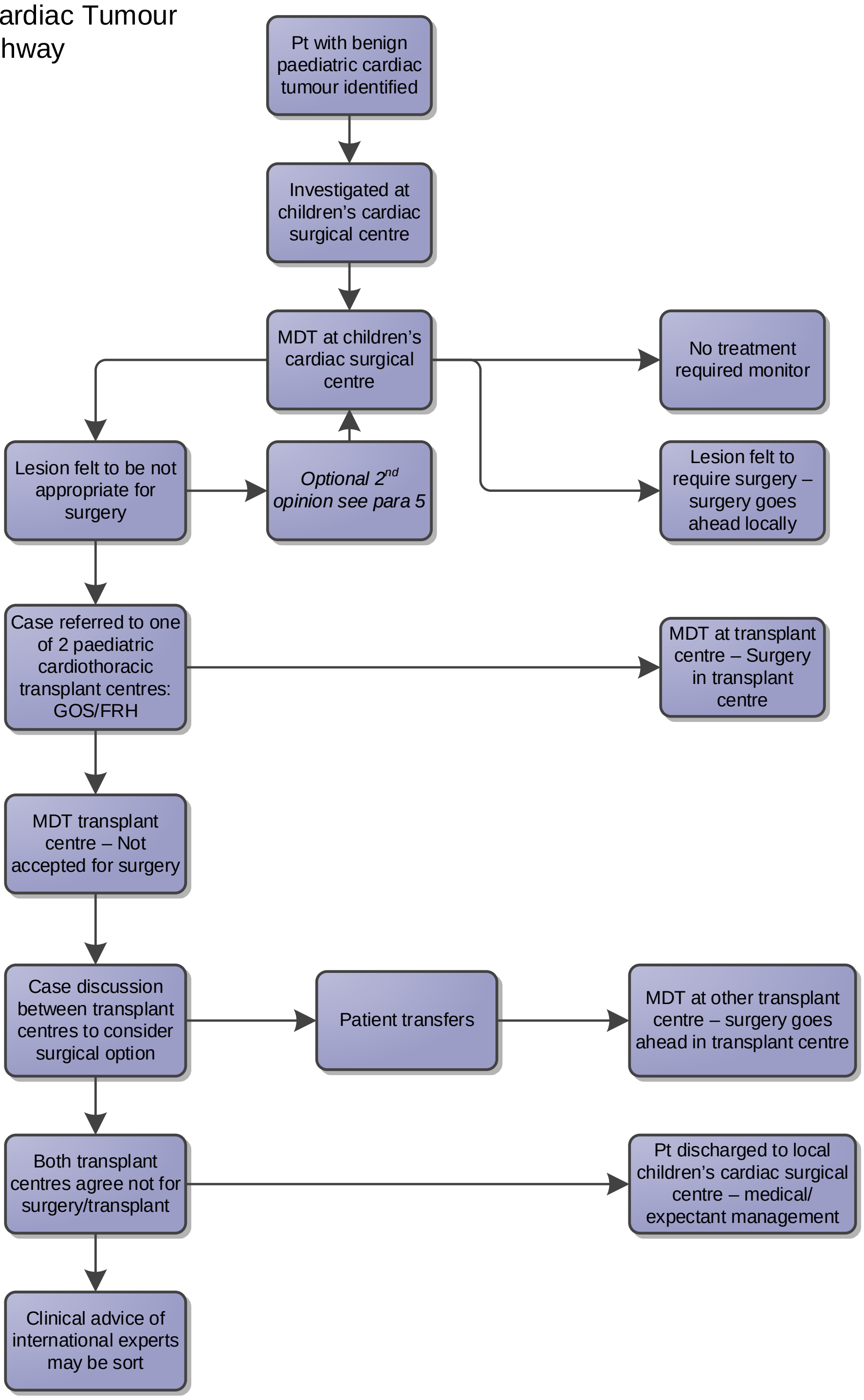
   ii. The transplant centres hold a joint discussion to agree if one centre is best placed to resect the tumour. That centre will then take over care of the patient.

Subject to the patient’s rights in law, NHS England will only consider funding a referral outside the UK for evaluation for possible treatment if:

a. this pathway has been followed and both UK paediatric cardiac transplant centres consider that the tumour is only resectable with subsequent transplant OR that the tumour is not resectable and the patient is ineligible for transplant AND
b. an overseas centre with relevant expertise and experience considers that resection without transplant is feasible AND
c. the referral is made by a UK transplanting centre.

Referral for evaluation outside the UK should only be made by the referring UK transplant centre MDT where there is a realistic prospect of successful removal.

Paediatric Cardiac Tumour Pathway



Effective from

11 July 2019

Recommendations for data collection

Data collection should include:

- Data on all patients entering the pathway to indicate their route through the algorithm e.g. at decision point 1 numbers assigned to ‘lesion felt to be not appropriate for surgery’, ‘lesion felt to require surgery – surgery goes ahead locally’, ‘no treatment required – monitor’ and similar for all other points of the pathway.

- 90-day, 1 and 5 year survival post operation for patients operated
- Time to significant events: use of ventricular assist devices, transplant
- Quality of life, using age appropriate instrument e.g. Peds QL, at age 2 and 5 years for all patients who enter the pathway
- Survival at age 1, 2 and 5 years old for all patients who enter the pathway

Mechanism for funding

NHS England will reimburse activity undertaken within the terms of this policy statement for patients treated in England under the tariffs for paediatric cardiac surgery.

Policy review date

This is an urgent policy statement, which means that the full process of policy production has been abridged: a full independent evidence review has not been conducted; and public consultation has not been undertaken. If a review is needed due to a new evidence base then a new Provisional Policy Proposal needs to be submitted by contacting england.CET@nhs.net.

Links to other Policies

None.

Equality Statement

Promoting equality and addressing health inequalities are at the heart of NHS England’s values. Throughout the development of the policies and processes cited in this document, we have:

- Given due regard to the need to eliminate discrimination, harassment and victimisation, to advance equality of opportunity, and to foster good relations between people who share a relevant protected characteristic (as cited under the Equality Act 2010) and those who do not share it; and

- Given regard to the need to reduce inequalities between patients in access to and outcomes from healthcare services and to ensure services are provided in an integrated way where this might reduce health inequalities.