



The Children &
Young People's
Cancer Association



child cancer
smart

A guideline to assist healthcare professionals in the assessment of children and young people who may have a soft tissue sarcoma

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Introduction

1.1 Background

Soft tissue sarcomas (STS) account for 7% of all childhood cancers^[1, 2]. The time to diagnosis (TTD) of STS is one of the longest of all childhood cancers^[3]. In the UK, the median TTD of STS is 6.9 weeks, but can be up to 205 weeks^[4]. There is a clear need to urgently review the clinical guidance available to healthcare professionals assessing those children and young people (CYP) who may have a soft tissue sarcoma.

The 2015 National Institute for Health and Care Excellence (NICE) published a guideline on ‘Suspected cancer: recognition and referral’ which covers all ages^[5]. There is a real need for paediatric-specific guidance, as adult cancers manifest and present differently. This NICE guidance is directed at primary care with the ‘end-point’ being referral onto secondary care. Children and young people with cancer experience diagnostic delay throughout the health service both at primary care and secondary care level, while a referral from primary to secondary care can add significant length to the patient’s diagnostic journey. Timely imaging is essential to identify children who need further investigations for soft tissue sarcomas and so guidance was required on indications for, and appropriate waiting times to ultrasound and/or MRI imaging.

Furthermore, the NICE recommendations lack a systematic evidence review and are based solely on expert consensus which notably did not include any paediatric oncologists. As a result, concern from the paediatric oncology community across the UK led to the publication of a supplement to the NICE guideline in 2021, following a Delphi consensus process conducted among the CCLG: The Children & Young People’s Cancer Association (CCLG) membership^[6, 7]. A full systematic evidence review was not completed at this time due to the urgent need for expert child-specific guidance to be published.

Detailed tumour-specific guidance is needed to empower clinicians to make decisions about those who need investigation and accelerate referrals for CYP with high-suspicion of soft tissue sarcoma promoting earliest possible diagnosis.

1.2 Aim of the guideline

This guideline aims to shorten the time to diagnosis of soft tissue sarcomas by providing evidence-based guidance for health professionals in primary and secondary care on the following:

1. The signs and symptoms that may occur in children and young people (CYP) with these tumours.
2. Assessment of CYP presenting with these signs and symptoms.
3. Indications and waiting times for imaging CYP with these signs and symptoms.

1.3 Clinical health questions

The guideline was devised to address the following questions:

1. What are the signs and symptoms that CYP with STS present with?
2. Given that the initial signs and symptoms of an STS may occur with other less serious childhood conditions, how can healthcare professionals distinguish those CYP who may have an STS from the majority who do not?

3. What is the best way to clinically assess a CYP presenting with these signs and/or symptoms?
4. What signs and/or symptoms in CYP increase the likelihood of an STS to the extent that their presence mandates imaging?
5. What is the best modality for imaging in these CYP?
6. In a CYP who presents with these signs and/or symptoms, what is an appropriate maximum waiting time to imaging?
7. Are there specific presentations of STS in CYP that are repeatedly associated with diagnostic difficulty?
8. Are there other barriers to diagnosis in STS in CYP and if so, how can these be addressed?

1.4 Scope and target population

The guideline is applicable to all children and young people aged 0-18 years who present with signs and/or symptoms that could be due to an STS and are being reviewed by a healthcare professional.

This guideline has been developed following the gold standard AGREE-II criteria. It reflected careful consideration of the available evidence and incorporated professional expertise via a Delphi consensus process. The levels of evidence and recommendation grades follow the framework developed by the Scottish Intercollegiate Guideline Network (SIGN)⁸.

The recommendations have also been summarised into a quick reference guide and a one-page poster for healthcare professionals (Appendix 1).

Guideline users

The guideline is intended to support all healthcare professionals caring for children and young people aged 0-18 years in their clinical practice.

Healthcare professionals should use it to support their decision-making when assessing children who may have an STS. It does not, however, override the responsibility of a healthcare professional to make decisions appropriate to the condition of individual children.

Stakeholder involvement

This guideline was developed under the leadership of CCLG: The Children & Young People's Cancer Association (CCLG) and The University of Nottingham. Key stakeholders, including general paediatricians, GPs, community paediatricians, emergency paediatricians, oncologists, paediatric surgeons and parent representatives with experience of childhood cancer diagnosis, volunteered to participate in the multi-disciplinary workshop and helped revise the statements following feedback from the Delphi panel.

The following organisations are also recognised as key stakeholders. While these organisations were not directly involved in the development of this guideline, they represent key contributors to the understanding, diagnosis, and management of paediatric STS:

- The National Institute for Health and Care Excellence (NICE)
- Scottish Intercollegiate Guidelines Network (SIGN)
- Royal College of Paediatrics and Child Health (RCPCH)
- Royal College of General Practitioners (RCGP)
- Royal College of Surgeons (RCS)

1.5 Funding

The guideline development has been funded by CCLG: The Children & Young People's Cancer Association, as part of the Child Cancer Smart awareness campaign.

1.6 Conflicts of interest

All GDG members, multidisciplinary workshop participants and Delphi consensus group participants declared no conflict of interests. The funders had no role in the guideline development.

Methods

2.1 Guideline development

The guideline was developed in accordance with the AGREE II criteria, following a three-stage process involving evidence review, expert consultation, and consensus building.

Figure 1

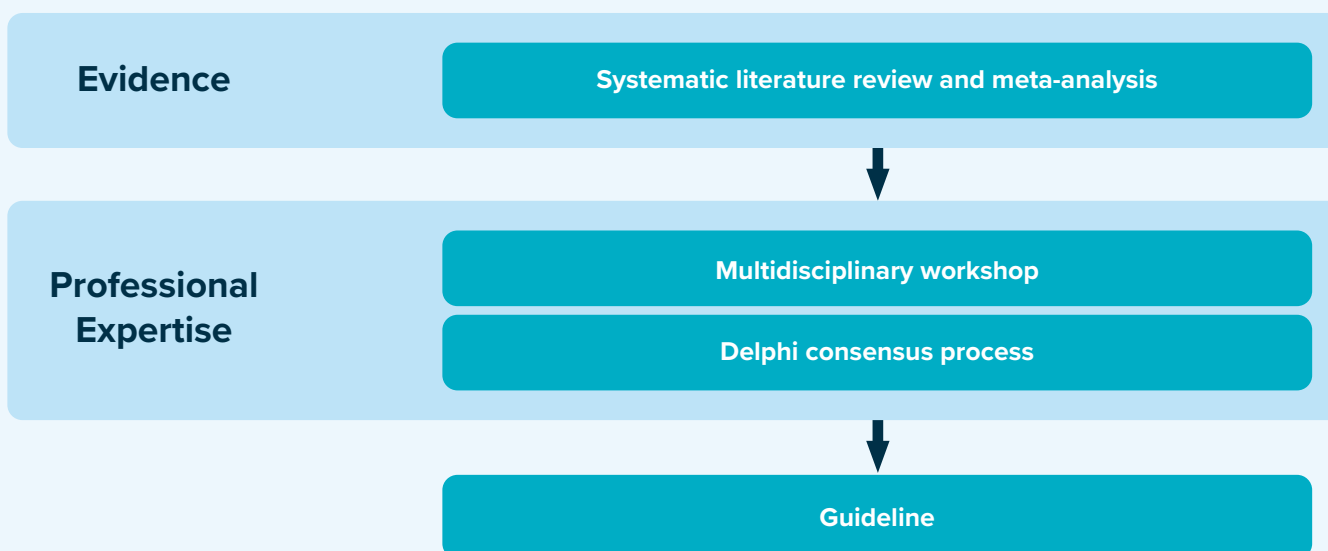


Figure 1. Guideline development methodology

The initial stage comprised a systematic review and meta-analysis to appraise current evidence on the clinical presentation of childhood soft tissue sarcoma. In stage two, a multidisciplinary workshop, involving clinicians from primary, secondary and tertiary care, as well as parents of children with cancer, reviewed the evidence and devised statements describing the clinical presentation, assessment, and investigation process. Parents were also given dedicated time to share their views. In stage three, these statements were refined through a modified Delphi process involving clinicians from primary, secondary, and tertiary care across relevant specialties. Membership of the Guideline Development Group (GDG), workshop, and Delphi panel is listed in Appendix 2.

2.2 Systematic review and meta-analysis

Clinical Question:

What are the signs and symptoms that children with soft tissue sarcomas develop?

A systematic review and meta-analysis of the presenting signs and symptoms of soft tissue sarcomas in children and young people under the age of 18 was carried out to provide the evidence base for the development of this guideline. The detailed methodology and full results including sub-analysis by tumour type have been published^[9].

2.3 Multidisciplinary workshop

Following the systematic review and meta-analysis, professional expertise was incorporated into guideline development to determine the specificity of signs and symptoms associated with childhood soft tissue sarcomas and to advise on appropriate referral pathways, imaging indications and acceptable waiting times.

Multidisciplinary workshop

The panel reviewed the data from the meta-analysis and initially discussed the presenting symptom list for soft tissue sarcomas, splitting each list into core symptoms and associated symptoms.

For each of the symptoms, the group was asked to devise statements on the following:

- How would the signs and symptoms present to a healthcare professional?
- How should a healthcare professional assess a child presenting with this sign or symptom?
- How should a healthcare professional determine whether the presenting signs and/or symptoms could be due to a soft tissue sarcoma, i.e. their specificity?
- What factors influence the specificity of a sign and/or symptom?
- What are appropriate thresholds for referral and selection for imaging for a child presenting with this sign and/or symptom?
- What would they regard as best practice for referral and imaging of a child presenting with this sign and/or symptom?

All discussions were translated into a series of statements by the GDG at the end of the workshop and sent back to the workshop participants to ensure they reflected the discussion.

2.4 Delphi consensus process

Statements for the first round of the Delphi consensus process were derived from the statements developed by the multidisciplinary workshop and from the evidence base provided by the systematic review. The detailed methodology and results have been published^[10].

Invitations to join the Delphi panel were sent to health specialists fulfilling one or more of the following criteria (Delphi panel composition - see Appendix 2):

- CCLG member from one of the following disciplines: paediatric surgery, paediatric radiology, paediatric oncology.
- A range of general practitioners, paediatric emergency physicians and paediatricians across the UK.

2.5 Strength of evidence and recommendation

The levels of evidence and grades of recommendations (Appendix 3) are based on the SIGN 50: A Guideline Developer's Handbook^[8]. Recommended best practice is based on the clinical experience of the GDG.

The Evidence

3.1 Systematic review and meta-analysis

The initial search of MEDLINE and EMBASE identified 11,115 titles published between January 2010 to February 2021, with no language restrictions. After title and abstract screening, 290 were selected for full-text review and 59 studies included in the final analysis^[11-69]. These studies provided data on 2462 cases of childhood soft tissue sarcomas, across 32 different countries.

The quality was comprehensively evaluated and summarised. A total of 101 symptoms/signs were recorded and overlapping clinical features were clustered together into **43 signs and symptoms**. Those with pooled proportions of 2% or more of the studied population are reported in Figures 2-4.

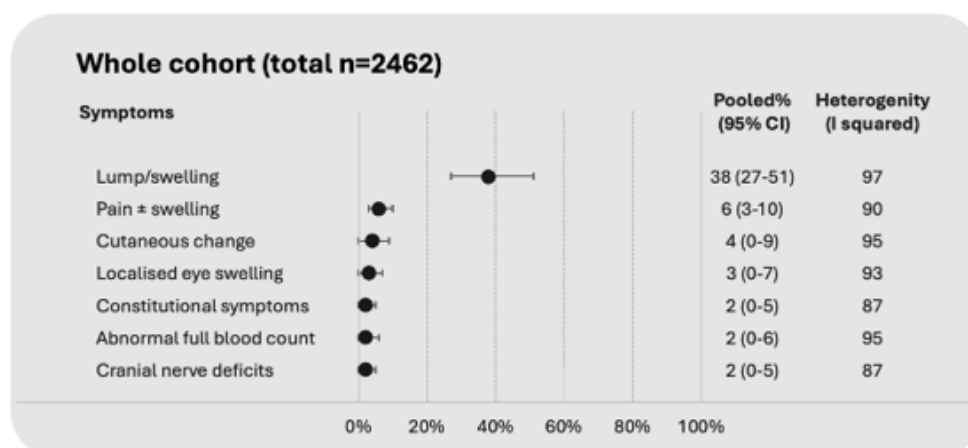


Figure 2.

Pooled proportions for the most frequently reported pre-diagnostic signs and/or symptoms in the whole cohort.

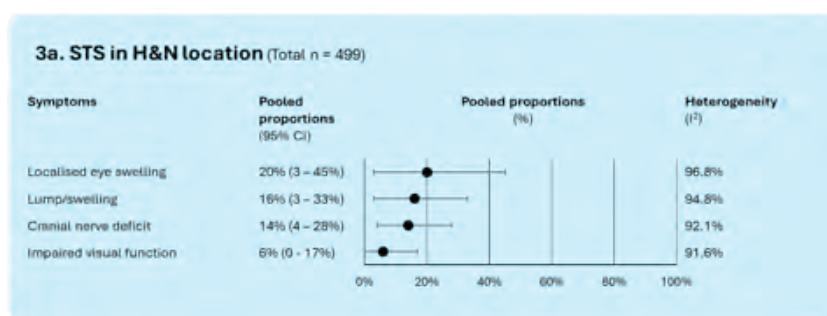
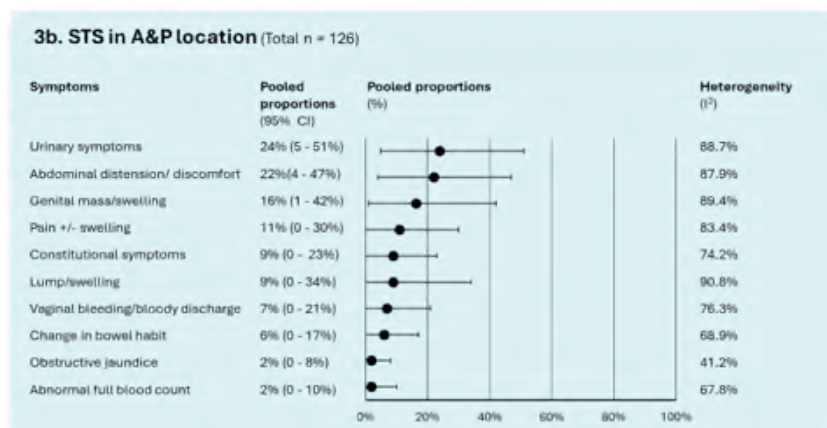


Figure 3.

Pooled proportions for the most common pre-diagnostic signs and/or symptoms reported for soft tissue sarcomas (STS) in (a) head and neck (H&N) and (b) abdomen and/or pelvis (A&P) locations



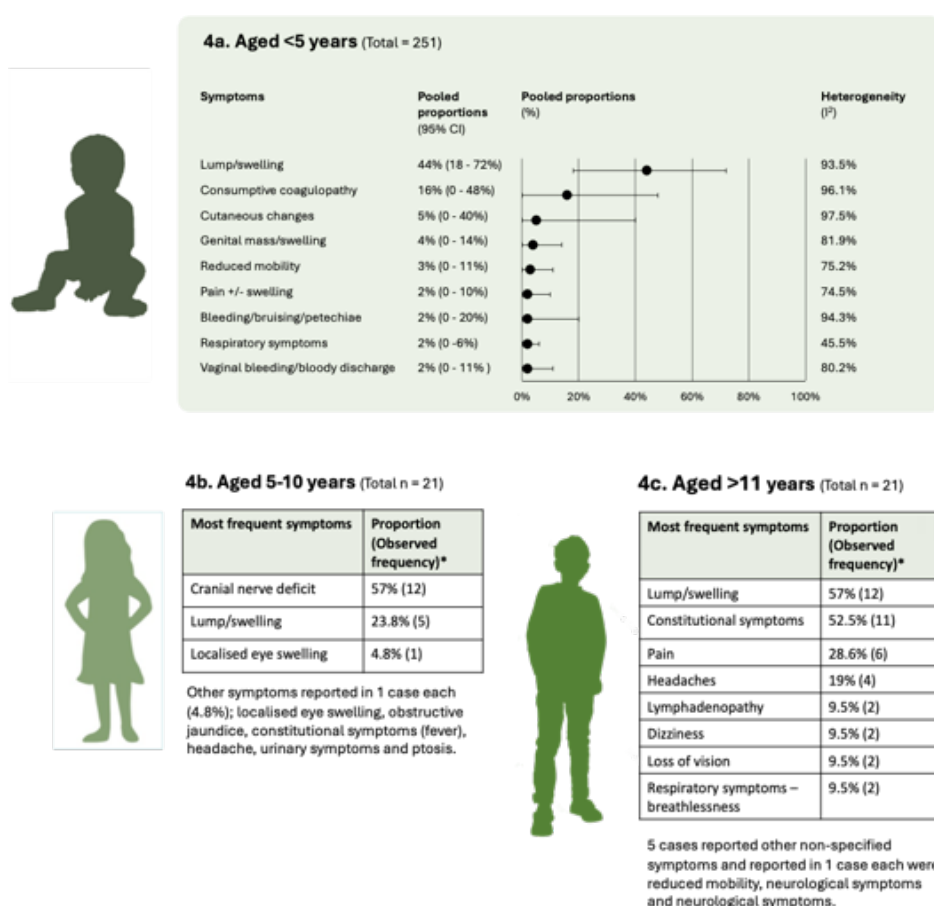


Figure 4.

Pre-diagnostic signs and/or symptoms reported for soft tissue sarcoma (STS) by age group. Pooled proportions for (a) ages <5 years, and proportions based on observed frequencies for (b) ages 5–10 years and (c) ages >11 years, where meta-analysis was not conducted due to the limited number of eligible papers. *Only two papers reported symptoms in ages 5–10 years and one paper in ages >11 years, therefore no meta-analysis was carried out in either category.

3.2 Core and associated symptoms

Soft tissue sarcomas can occur anywhere in the body, and signs and symptoms can differ depending on where the sarcoma originates. Soft tissue sarcoma presenting symptoms can be grouped into limb, thoracic, head and neck and abdomen and pelvis.

A systematic review and meta-analysis showed that soft tissue sarcomas in children and young people present with the following core and associated presenting symptoms^[9].

Core symptoms:

The following signs and symptoms are all associated with childhood soft tissue sarcomas. Their presence should alert the clinician to this possibility.

- Unexplained lump
- Swelling
- Pain
- Skin changes over the lump/swelling

Associated symptoms:

- Limbs: lump, limp, restricted movement or altered function
- Head and neck: localised eye swelling, cranial nerve deficit, impaired vision, ear swelling or pain.
- Systemic symptoms: weight loss, fever, malaise
- Abdomen and pelvis: urinary symptoms, abdominal/flank distension, genital lump/swelling, vaginal bleeding, change in bowel habit, obstructive jaundice

- Bruising/bleeding, especially in CYP aged under one year old
- Back pain (neck to sacrum) with or without signs of spinal cord compression

3.3 Delphi consensus

The Delphi consensus process comprised of 48 statements describing best practice (referral, imaging, predisposing factors), the presenting features of childhood soft tissue sarcomas (general, unexplained lump, swelling, pain, skin changes, limp and restricted movement/altered limb function), factors that could be used to discriminate these tumours from other less serious conditions and possible referral pathways for children with signs or symptoms suggestive of soft tissue sarcomas.

The initial round achieved a consensus in 47 out of 48 statements sent out for review and all 48 statements reached consensus within two rounds. The full Delphi consensus process methodology and results for best practice and soft tissue sarcoma statements have been published^[10].

Table 1 - Best practice statements and percentages of consensus

Section A: Best practice (statements 1-12)			
Consultation	1	Explicitly ask young people, parents and carers about their concerns regarding what the cause of their symptoms are in any consultation.	92%
	2	If a parent/carer expresses concerns about a possible tumour this should be reviewed carefully. If a tumour is unlikely, explain why and give appropriate safety netting advice.	96%
	3	Any healthcare professional deciding to review a patient to diagnose or exclude cancer should consider the risk of rapid tumour growth. They must ensure that the timing of review does not exceed the national 28 day limit to access a diagnostic test and obtain the result.	83%
	4	Offer a telephone or in-person interpreter if the patient, parent / carer, or healthcare professional are not fluent in English or Welsh.	94%
	5	Be aware that low parental educational level, social deprivation and lack of familiarity with the UK healthcare system may be associated with diagnostic delay. Consider a multi-disciplinary approach for these families (for example health visitor liaison) and provide clear written safety netting for when to seek further medical advice.	87%
	6	Be aware that the presence of complex neuro-disabilities or other communication difficulties (e.g. Autistic Spectrum Disorder) may also be associated with diagnostic delay. Care should be taken to elicit concerns from parents or carers that know them best.	92%
Referral	7	In primary care, discuss concerns with your local consultant paediatrician on-call on the same day if there is a high index of suspicion (i.e. "I think this child has cancer") regarding a possible soft tissue sarcoma in a child/young person (CYP).	86%

	8	In primary care, discuss concerns with the local consultant paediatrician on-call before referring a CYP* from primary care in which the differential diagnosis includes a possible soft tissue sarcoma (“I am not sure what this is, but there is a possibility it could be cancer”) to ensure the CYP* is seen within the most suitable timeframe.	79%
	9	Be aware that soft tissue sarcomas can progress rapidly over days, increasing in size and causing pressure effects (e.g. breathing difficulties or spinal cord compression). If there is suspicion of a rapidly growing soft tissue sarcoma this should be discussed with a paediatrician to decide the most appropriate time for review, which will often be the same day.	92%
Imaging	10	Be aware that ultrasound imaging is not always the most suitable imaging modality for a soft tissue sarcoma. In secondary care, discuss with a paediatric radiologist to decide upon the most suitable imaging of choice.	80%
	11	When requesting imaging, write explicitly in the request that a soft tissue sarcoma is suspected.	87%
Predisposing factors	12	Be aware that some predisposing factors such as history of Neurofibromatosis 1, Li-Fraumeni syndrome, DICER1 or family history of breast or ovarian cancer at a young age or a sibling with childhood cancer, are associated with an increased risk of soft tissue sarcomas. Verify the presence of predisposing factors with parents/patients as they may lower the threshold for referral and investigation.	85%

Table 2 - Soft tissue sarcoma statements and percentages consensus

Section B: Soft tissue sarcomas statements (Statements 13-48)			
General	13	Take a detailed history, including the presence or absence of the other symptoms on the list above, predisposing factors and a family history for CYP* presenting with symptoms suggestive of a soft tissue sarcoma.	93%
	14	Be aware that soft tissue sarcomas can present with systemic symptoms such as fever, malaise and weight loss. Ask about these symptoms when seeing patients with other symptoms suggestive of a soft tissue tumour.	95%
	15	Be aware that soft tissue sarcomas causing spinal cord compression can affect bladder, bowel and erectile dysfunction. Ask for the presence of urinary or faecal incontinence and erectile dysfunction when taking a history, especially if the presenting complaint is back pain.	94%
	16	Examination of the lump/swelling should include a regional lymph node assessment.	94%
	17	Examination of the child will be guided by presenting symptoms; however, it should include examination of the skin (birth marks e.g. café au lait spots) and a neurological examination.	79%
	18	In secondary care, measure and plot weight measurements on age-appropriate growth charts for CYP* with signs or symptoms suggestive of a soft tissue sarcoma.	77%
	19	Be aware that if symptoms or clinical suspicion persists despite normal initial imaging, a discussion with the radiologist about the most appropriate repeat imaging is advised.	79%
Unexplained lump	20	Ask about the presence of the other symptoms of a soft tissue sarcoma (swelling, pain, skin changes, fever, weight loss, back pain, bowel/bladder/erectile dysfunction, birth marks e.g. café au lait spots) in a CYP* presenting with an unexplained lump.	88%
	21	Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain and swelling or hearing loss) if the lump is in the head/neck area.	89%
	22	Ask about the presence of new onset of swallowing difficulties / choking, stridor or snoring, the presence of new onset of respiratory symptoms.	85%
	23	Ask about the presence of abdomen/pelvic symptoms (abdominal distension, genital mass, vaginal bleeding) if the lump originates in the abdomen/pelvis.	92%
	24	In primary care, request urgent review by paediatrics for any unexplained persistent and growing (present for two weeks) lump.	84%
	25	In secondary care, request urgent imaging for any unexplained persistent and growing (present for two weeks) lump.	85%

	26	In primary care, request urgent review for rapidly increasing size of lump or increasing number of lumps.	87%
	27	In secondary care, request urgent imaging for any unexplained lump with another symptom from the symptom checklist.	83%
	28	Be aware that diagnostic delay has been associated with attributing lumps to lipomas or haemangiomas without typical appearance. If there is diagnostic uncertainty and no improvement, always discuss with paediatricians or paediatric surgeons for a second opinion or review.	92%
	29	Be aware that diagnostic delay has been associated with misdiagnosis of a benign mass on imaging, leading to an incorrect surgical procedure. Imaging should be reported by paediatric radiologists, and any diagnostic uncertainty should be discussed at the appropriate paediatric oncology MDT prior to surgery.	91%
Swelling	30	Ask about the presence of the other symptoms of a soft tissue sarcoma (lump, pain, skin changes, birth marks (café au lait spots), fever, weight loss, back pain and bowel/bladder/erectile dysfunction) in a CYP* presenting with persistent or progressing swelling (present for a 2-week period or more).	88%
	31	Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain or swelling or localised hearing loss) if the lump is in the head/neck area.	89%
	32	Be aware that delayed diagnosis has been associated with attributing a red warm swelling to infection despite no improvement with antibiotics. Arrange to see the CYP* at the end of the course of antibiotics to assess response. If there has been no response, consider discussion with secondary care for advice or referral.	87%
	33	In primary care, request an urgent review for any persistent swelling (present for two weeks or more), that is rapidly increasing in size, including eye or ear swelling.	87%
	34	In secondary care, request imaging for any persistent swelling (present for two weeks or more) that is rapidly increasing in size, including eye or ear swelling.	85%
	35	In primary care, request an urgent review for any persistent swelling (present for two weeks or more) not resolving despite treatment with regular anti-inflammatories OR antibiotics.	79%
	36	In secondary care, request imaging for any persistent swelling (present for two weeks or more) not resolving despite treatment with regular anti-inflammatories OR antibiotics.	81%
Pain	37	Be aware that children aged younger than four years, or those with communication difficulties, are frequently unable to describe or localise pain; their behaviour e.g. withdrawal, holding their leg, not weight bearing may indicate bone pain. Look for these signs on examination.	95%
	38	Ask and examine for the other signs and symptoms suggestive of a soft tissue sarcoma (lump, swelling, skin changes, birth marks (café au lait), fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present) in CYP* with pain.	88%

	39	In primary care, request an urgent review for any painful, persistent (present for two weeks or more) or progressive lump or swelling.	83%
	40	In secondary care, request imaging for any painful, persistent (present for two weeks or more) or progressive lump or swelling.	86%
Skin changes over the lump/swelling	41	Ask about the presence of the other symptoms of a soft tissue sarcoma in a CYP* presenting with skin changes over a lump or swelling pain (fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present).	92%
	42	Be aware that soft tissue sarcomas in under-ones have a risk of consumptive coagulopathy (Kasabach Merrit syndrome). A full blood count is recommended in children under one with a suspected soft tissue sarcoma, especially if presenting with bruising and petechiae.	81%
	43	In primary care, request an urgent review for a CYP* who has a persisting or progressive lump/swelling (for two weeks) with overlying skin changes.	84%
	44	In secondary care, request imaging for a CYP* who has a persisting or progressive lump/swelling (for two weeks) with overlying skin changes.	88%
Associated symptoms: Limp, restricted movement or altered limb function	45	Have a high level of concern for a CYP* who is normally highly active or sporty but is no longer able to play sport due to the unexplained limb pain or altered limb function.	95%
	46	Be aware that abdominal masses can cause neurological symptoms due to pressure on the spinal cord. The pressure can cause children to present as “off legs” or refusal to weight bear. Examine the abdomen and spine when a CYP* presents with refusal to weight bear.	94%
	47	In primary care, request an urgent review for a CYP* who is non-weight bearing or has restricted movement or altered limb function.	87%
	48	In secondary care, request imaging for a CYP* who is non-weight bearing or has restricted movement despite adequate analgesia.	84%

*CYP, children and young people.

Clinical Guideline

4.1 Best practice – consultation

Explicitly ask young people, parents and carers about their concerns regarding what the cause of their symptoms are in any consultation.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 92% (Delphi round 1)
- Rationale: Parents / carers of children with persistent symptoms are frequently concerned that their child's symptoms may be due to a tumour for a significant period of time before the diagnosis is made. Parents / carers may be unwilling to express these concerns for fear of seeming overly anxious or appearing to waste healthcare professionals' time. Explicitly asking parents / carers of their concerns enables them to be expressed, improving communication between all parties. In some cases, parental concern regarding a possible tumour may trigger professional concern and lead to appropriate investigation.

If a parent/carer expresses concerns about a possible tumour this should be reviewed carefully. If a tumour is unlikely, explain why and give appropriate safety netting advice.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 96% (Delphi round 1)
- Rationale: Parents / carers of children with soft tissue sarcomas are frequently concerned that their child's symptoms may indicate a tumour for a significant period of time before the diagnosis is made. If, on review, a tumour seems unlikely it is important to explain why, in order to maintain trust and communication with the patient and their parents / carers.

Any healthcare professional deciding to review a patient to diagnose or exclude cancer should consider the risk of rapid tumour growth. They must ensure that the timing of review does not exceed the national 28 day limit to access a diagnostic test and obtain the result.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 83% (Delphi round 1)
- Rationale: Symptom progression occurs with childhood cancers; therefore early review is recommended to facilitate detection of any associated symptoms or signs which may make the diagnosis more likely. The current Faster Diagnosis Standard aims for all patients with suspected cancer to have a diagnosis or the “all clear” within 28 days and the Guideline Development Group (GDG) felt that this target should be reflected in this guideline^[70].

Offer a telephone or in person interpreter if the patient, parent / carer or healthcare professional are not fluent in English or Welsh.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 94% (Delphi round 1)
- Rationale: The Guideline Development Group (GDG), multi-disciplinary workshop and Delphi panel could all identify individual cases where non-English first language was associated with diagnostic delay. It is essential to take a thorough history when assessing a child who may have a tumour; this is not possible if the patient, parent/carers and healthcare professional are not fluent in a common language.

Be aware that low parental educational level, social deprivation and lack of familiarity with the UK healthcare system may be associated with diagnostic delay. Consider a multi-disciplinary approach for these families (for example health visitor liaison) and provide clear written safety netting for when to seek further medical advice.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 87% (Delphi round 1)
- Rationale: There is no published evidence linking low parental education, social deprivation and lack of familiarity with the UK healthcare system with diagnostic delay in soft tissue sarcomas. However, the guideline development team and many members of the first Delphi panel were aware of individual cases in which these factors may have contributed to a prolonged symptom interval. These children may not necessarily need quicker referral but would benefit with thorough safety netting and health visitor support to ensure any new symptoms are not missed.

Be aware that the presence of complex neuro-disabilities or other communication difficulties (e.g. Autistic Spectrum Disorder) may also be associated with diagnostic delay. Care should be taken to elicit concerns from parents or carers that know them best.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 92% (Delphi round 1)
- Rationale: The multi-disciplinary workshop team highlighted that those with complex neuro-disabilities or other communication difficulties were also more likely to experience a diagnostic delay. Parents may only note a change in their behaviour without knowing the reason why. Care should be taken to listen to parental concern.

4.2 Best practice – referral

In primary care, discuss concerns with your local consultant paediatrician on-call on the same day if there is a high index of suspicion (i.e. “I think this child has cancer”) regarding a possible soft tissue sarcoma in a child/young person (CYP).

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 86% (Delphi round 1)
- Rationale: Children with tumours may deteriorate quickly. Therefore, if there is a high likelihood that a child may have a tumour, they should be assessed promptly and arrangements for imaging should be made as quickly as possible.

Be aware that soft tissue sarcomas can progress rapidly over days, increasing in size and causing pressure effects (e.g. breathing difficulties or spinal cord compression). If there is suspicion of a rapidly-growing soft tissue sarcoma this should be discussed with a paediatrician to decide the most appropriate time for review, which will often be the same day.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 92% (Delphi round 1)
- Rationale: The Guideline Development Group (GDG) and multi-disciplinary workshop members felt that there was low awareness of how soft tissue sarcomas can progress rapidly and the urgency with which these children and young people need to be seen to ensure prompt diagnosis.

In primary care, discuss concerns with the local consultant paediatrician on-call before referring a CYP from primary care in which the differential diagnosis includes a possible soft tissue sarcoma (“I am not sure what this is, but there is a possibility it could be cancer”) to ensure the CYP is seen within the most suitable timeframe.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 79% (Delphi round 1)
- Rationale: A prolonged symptom interval with soft tissue sarcomas occurs in part due to delay between initial referral from primary care and assessment in secondary care. The Department of Health has advised that a patient (adult or child) presenting with symptoms that are potentially indicative of a malignancy should be assessed by a healthcare professional with expertise in that area within two weeks^[71]. However, data shows that only 2% of children referred via the old two week wait system (now known as urgent suspected cancer pathway) actually receive a childhood cancer diagnosis^[72,73]. The paediatricians amongst the multi-disciplinary workshop felt strongly that cases should be discussed with them if there is a suspicion of cancer as they would be able to ensure the child or young person is seen in the most appropriate place as soon as possible.

4.3 Best practice - imaging

Soft tissue sarcomas can occur anywhere in the body. Imaging of lumps/suspected sarcomas should be guided by paediatric radiologists, and so imaging is to be requested once the CYP has been seen in secondary care.

Be aware that ultrasound imaging is not always the most suitable imaging modality for a soft tissue sarcoma. In secondary care, discuss with a paediatric radiologist to decide upon the most suitable imaging of choice.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 80% (Delphi round 1)
- Rationale: Early discussion with a paediatric radiologist helps to ensure the most appropriate imaging modality for the specific location of lump/suspected sarcoma is arranged to gain sufficient clinical information to guide early diagnosis and minimise potential delays to diagnosis from unsuitable imaging. For example, a soft tissue sarcoma in the head and neck region may need an MRI, whilst a peripheral soft tissue sarcoma may need an X-ray and an ultrasound.

When requesting imaging, write explicitly in the request that a soft tissue sarcoma is suspected.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 87% (Delphi round 1)
- Rationale Normal and abnormal imaging findings can vary significantly between children and adults. In order to reduce the risk of misdiagnosis, the Delphi panel agreed that soft tissue sarcoma should be mentioned as a potential differential diagnosis to guide interpretation and that imaging in children should be interpreted by a healthcare professional with expertise in this area.

4.4 Predisposing factors

Be aware that some predisposing factors such as history of Neurofibromatosis 1, Li-Fraumeni syndrome, DICER1 or family history of breast or ovarian cancer at a young age or a sibling with childhood cancer, are associated with an increased risk of soft tissue sarcomas. Verify the presence of predisposing factors with parents/patients as they may lower the threshold for referral and investigation.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 85% (Delphi round 1)
- Rationale: The above are all associated with an increased risk of childhood soft tissue sarcomas and therefore their presence should alert the clinician to this possibility and may lower their threshold for referral and investigation^[74].

4.5 Presentation of a child with a potential soft tissue sarcoma

Soft tissue sarcomas can occur anywhere in the body, and signs and symptoms can differ depending on where the sarcoma originates.

Soft tissue sarcoma symptoms can be grouped into limb, thoracic, head and neck and abdomen/pelvis.

A systematic review and meta-analysis showed that soft tissue sarcomas in children and young people present with the following core and associated symptoms.

Presenting signs and symptoms

The following signs and symptoms are all associated with childhood soft tissue sarcomas. Their presence should alert the clinician to this possibility.

- Unexplained lump
- Swelling

- Pain
- Skin changes over the lump/swelling

Associated symptoms*:

- Limbs: lump, limp, restricted movement or altered limb function
- Head and neck: localised eye swelling, cranial nerve deficit, impaired vision, ear swelling or pain
- Abdomen and pelvis: urinary symptoms, abdominal/flank distension, genital lump/swelling, vaginal bleeding, change in bowel habit, obstructive jaundice
- Systemic symptoms: weight loss, fever, malaise
- Bruising/ bleeding, especially in CYP aged under one year
- Back pain (neck to sacrum) with or without signs of spinal cord compression

**Associated symptoms are dependent on the type and location of the tumour and it is important to ask for the presence or absence of these symptoms.*

The selection of core and associated symptoms was based on data from meta-analyses and discussions at a multidisciplinary workshop. The Guideline Development Group (GDG) and the multi-disciplinary workshop members agreed on the symptoms which are categorised as associated symptoms. Whilst the associated symptoms are not individually common, the multidisciplinary team members all agreed that in combination with the core symptoms, these associated symptoms should warrant consideration of soft tissue sarcomas.

Abdominal soft tissue sarcoma signs and symptoms can overlap with the signs and symptoms of bone and other abdominal tumours. There are already published guidelines and decision support tools for childhood bone and abdominal tumours. Therefore, it was agreed by the GDG and multidisciplinary workshop members that limp, restricted movement or altered limb function should be included as associated symptoms to be aware of. Please see bone (<https://www.cclg.org.uk/child-cancer-smart/bone-tumours>) and/or abdominal tumour (<https://www.cclg.org.uk/childcancersmart/resources-for-professionals/abdominal-tumours>) decision support tool for CYP presenting with a limp, restricted movement or altered limb function who could have a tumour.

4.6 Assessment of a child with a potential soft tissue sarcoma

4.6.1 History

Take a detailed history, including the presence or absence of the other symptoms on the list above, predisposing factors and a family history for CYP presenting with symptoms suggestive of a soft tissue sarcoma.

- | | |
|------------------------|---|
| • Strength of evidence | 4 |
| • Recommendation form | Strong |
| • Consensus achieved | 93% (Delphi round 1) |
| • Rationale: | Taking a detailed history including specifically enquiring about any other symptoms and/or predisposing factors helps to identify those children who may have tumours and need imaging from the majority who don't. |

4.6.2 Assessment

Be aware that soft tissue sarcomas can present with systemic symptoms such as fever, malaise and weight loss. Ask about these symptoms when seeing patients with other symptoms suggestive of a soft tissue tumour.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 95% (Delphi round 1)
- Rationale: Often CYP, especially teenagers, presenting with an unexplained lump do not recognise systemic symptoms as relevant, therefore do not automatically mention these during consultations. The GDG and multidisciplinary team felt that it is important to specifically ask about such symptoms when CYP present with an unexplained lump to guide clinical suspicion of a soft tissue sarcoma.

Be aware that soft tissue sarcomas causing spinal cord compression can affect bladder, bowel and erectile dysfunction. Ask for the presence of urinary or faecal incontinence and erectile dysfunction when taking a history, especially if the presenting complaint is back pain.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 94% (Delphi round 1)
- Rationale: Although this presentation is uncommon, the GDG and multidisciplinary team agreed that prompt recognition of such symptoms from spinal cord compression is essential to guide urgent review and intervention.

Examination of the lump/swelling should include a regional lymph node assessment.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 94% (Delphi round 1)
- Rationale: The multidisciplinary team agreed that assessment of regional lymph nodes is a necessary part of any thorough review of a CYP presenting with an unexplained lump/swelling.

Examination of the child will be guided by presenting symptoms; however, it should include examination of the skin (birth marks, e.g. café au lait spots) and a neurological examination.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 79% (Delphi round 1)
- Rationale: The multidisciplinary team felt that when healthcare professionals are assessing a CYP presenting with symptoms that could be a soft tissue sarcoma they should perform a skin and neurological examination to identify any clinical signs that may increase the risk of soft tissue sarcoma and guide clinical suspicion.

In primary care, ask about weight loss in CYP with signs or symptoms suggestive of a soft tissue sarcoma. If possible, weigh the CYP and compare with previous weights or plot on a growth chart.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved Not applicable
- Rationale: This statement was agreed by the multidisciplinary team and Guideline Development Group (GDG) as important and relevant to the assessment of any CYP with symptoms that could be a soft tissue sarcoma. This statement did not reach numerical consensus. However, while respondents agreed this was important, they felt that it was not always possible to perform in primary care and so this is phrased as 'if possible'. Healthcare professionals should be aware that weight loss is not always present, and its absence should not provide false reassurance or delay referral to secondary care if clinical suspicion is high.

In secondary care, measure and plot weight measurements on age-appropriate growth charts for CYP with signs or symptoms suggestive of a soft tissue sarcoma.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 77% (Delphi round 2)
- Rationale: The multidisciplinary team and GDG agreed this should be done for all CYP seen in secondary care with a suspected soft tissue sarcoma. Healthcare professionals should be aware that weight loss is not always present, and its absence should not provide false reassurance or delay referral to secondary care if clinical suspicion is high.

Be aware that if symptoms or clinical suspicion persists despite normal initial imaging, a discussion with the radiologist about the most appropriate repeat imaging is advised.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 79% (Delphi round 1)
- Rationale: Imaging findings can be difficult to interpret, and findings can progress over time, highlighting the importance of gaining the most appropriate imaging modality. A common pitfall leading to delays in diagnosis occurs when imaging suggests a benign aetiology. If clinical suspicion remains high and symptoms continue to progress despite 'reassuring' initial imaging, discussion with a radiologist who has expertise in this area is essential to arranging the most appropriate repeat imaging modality and minimising delays to diagnosis.

4.7 Signs and symptoms of a child with a potential soft tissue sarcoma

4.7.1 Unexplained lump

Ask about the presence of the other symptoms of a soft tissue sarcoma (swelling, pain, skin changes, fever, weight loss, back pain, bowel/bladder/erectile dysfunction, birth marks, e.g café au lait spots) in a CYP presenting with an unexplained lump.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 88% (Delphi round 1)

Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain and swelling or hearing loss) if the lump is in the head/neck area.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 89% (Delphi round 1)

Ask about the presence of new onset of swallowing difficulties/choking, stridor or snoring, the presence of new onset of respiratory symptoms.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 85% (Delphi round 1)

Ask about the presence of abdomen/pelvis symptoms (abdominal distension, genital mass, vaginal bleeding) if the lump originates in the abdomen/pelvis.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 92% (Delphi round 1)

In primary care, request urgent review by paediatrics for any unexplained persistent and growing (present for 2 weeks) lump.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 84% (Delphi round 1)

In primary care, request urgent review for rapidly increasing size of lump or increasing number of lumps.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 87% (Delphi round 1)

Rationale:

The multidisciplinary team and GDG agreed on these statements as important for assessing a CYP with an unexplained lump and as such emphasised in these guidelines. It is particularly important to explore the presence of associated symptoms or progression of symptoms which can inform the urgency of referral and investigations.

Diagnostic pitfalls

Be aware delayed diagnosis has been associated with:

Attributing lump to an injury without a clear history

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved not applicable
- Rationale: The GDG and multi-disciplinary team felt that it was particularly important to highlight cases which, whilst not necessarily common, were, in their experience, particularly associated with a prolonged symptom interval and diagnostic difficulty. This statement was expert-initiated, not sent out for consensus.

Attributing lumps to lipomas or haemangiomas without typical appearance. If there is diagnostic uncertainty and no improvement, always discuss with a paediatrician or a paediatric surgeon for a second opinion or review.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 92% (Delphi round 1)
- Rationale: The GDG and multi-disciplinary team felt that it was particularly important to highlight cases which, whilst not necessarily common, were, in their experience, particularly associated with a prolonged symptom interval and diagnostic difficulty. In order to make these areas easy to identify in the guideline, they have been headed with the caption “Delayed diagnosis has been associated with:”.

Misdiagnosis of a benign mass on imaging, leading to an incorrect surgical procedure. Imaging should be reported by paediatric radiologists, and any diagnostic uncertainty should be discussed at the appropriate paediatric oncology MDT prior to surgery

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 91% (Delphi round 1)
- Rationale: The GDG and multi-disciplinary team felt that it was particularly important to highlight cases which, whilst not necessarily common, were, in their experience, particularly associated with a prolonged symptom interval and diagnostic difficulty. In order to make these areas easy to identify in the guideline, they have been headed with the caption “Delayed diagnosis has been associated with:”.

Requesting imaging for

In primary care, requesting imaging should not delay referral/discussion with paediatrics

In secondary care, request urgent imaging for any unexplained persistent* and growing lump

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 85% (Delphi round 1)

**persistent = present for two weeks or more*

In secondary care, request urgent imaging for unexplained lump with another symptom from the symptom checklist

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 83% (Delphi round 1)

4.7.2 Swelling

Ask about the presence of the other symptoms of a soft tissue sarcoma (lump, pain, skin changes, birth marks (café au lait spots), fever, weight loss, back pain and bowel/bladder/erectile dysfunction) in a CYP presenting with persistent or progressing swelling (present for a two-week period or more).

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 88% (Delphi round 1)

Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain or swelling or localised hearing loss) if the lump is in the head/neck area.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 89% (Delphi round 1)

In primary care, request an urgent review for any persistent swelling (present for two weeks or more) rapidly increasing in size, including eye or ear swelling.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 85% (Delphi round 1)

In primary care, request an urgent review for any persistent swelling (present for two weeks or more) not resolving despite treatment with regular anti-inflammatories OR antibiotics.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 79% (Delphi round 1)

Rationale:

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed on these statements for assessing a CYP with an unexplained swelling. It is particularly important to ascertain the presence of associated symptoms or progression of symptoms which help inform urgency of referral and investigations.

Diagnostic pitfalls

Be aware delayed diagnosis has been associated with:

Attributing a red warm swelling to infection despite no improvement with antibiotics. Arrange to see the CYP at the end of the course of antibiotics to assess response. If there has been no response, consider discussion with secondary care for advice or referral.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 87% (Delphi round 1)
- Rationale: The GDG and multi-disciplinary team felt that it was particularly important to highlight cases which, whilst not necessarily common, were, in their experience, particularly associated with a prolonged symptom interval and diagnostic difficulty. In order to make these areas easy to identify in the guideline, they have been headed with the caption “Delayed diagnosis has been associated with:”.

Requesting imaging for

In primary care, requesting imaging should not delay referral/discussion with paediatrics

In secondary care, request imaging for any persistent swelling* rapidly increasing in size, including an eye or ear swelling.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 87% (Delphi round 1)

In secondary care, request imaging for any persistent swelling* not resolving despite treatment with regular anti-inflammatories OR antibiotics.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 81% (Delphi round 1)

**persistent = present for two weeks or more*

4.7.3 Pain

Be aware that children aged younger than four years, or those with communication difficulties, are frequently unable to describe or localise pain; their behaviour e.g. withdrawal, holding their leg, not weight bearing may indicate bone pain. Look for these signs on examination.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 95% (Delphi round 1)

Ask and examine for the other signs and symptoms suggestive of a soft tissue sarcoma (lump, swelling, skin changes, birth marks (café au lait), fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present) in CYP with pain.

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 88% (Delphi round 1)

In primary care, request an urgent review for a painful, persistent or progressive (present for two weeks or more) lump or swelling.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 83% (Delphi round 1)

Rationale:

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed that while pain is a very common childhood presentation, in most cases it is not due to a tumour. Limping or refusing to weight bear is an unusual childhood presentation and a thorough examination is essential for any child presenting with persistent unexplained pain or who may not be able to communicate their pain. Each child with persistent pain should be assessed for the presence of any core symptoms and associated symptoms. The presence of associated symptoms can help inform urgency of referral and investigations.

Diagnostic pitfalls

Not performing a full examination for those presenting with pain that may be referred pain.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved Not applicable
- Rationale: The multidisciplinary team and GDG can recall this diagnostic pitfall being associated with delays to diagnosis. Therefore, the GDG and multidisciplinary workshop group agreed that this statement should be highlighted in the guideline. This statement is expert-initiated and was not sent out for consensus.

Requesting imaging for

(In primary care, requesting imaging should not delay referral/discussion with paediatrics)

In secondary care, request imaging for any painful, persistent* or progressive lump or swelling.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 86% (Delphi round 1)

**persistent = present for two weeks or more*

4.7.4 Skin changes over the lump/swelling

Ask about the presence of the other symptoms of a soft tissue sarcoma in a CYP presenting with skin changes over a lump or swelling pain (fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present).

- Strength of evidence 2++
- Recommendation form Strong
- Consensus achieved 92% (Delphi round 1)

In primary care, request an urgent review for a CYP who has a persisting or progressive lump/swelling (for two weeks) with overlying skin changes.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 84% (Delphi round 1)

Rationale:

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed on these statements. The identification of progressive symptoms, development of associated symptoms at any of these reviews, the presence of diagnostic uncertainty or the failure to improve despite seemingly appropriate treatment should be referred to secondary care.

Diagnostic pitfalls

Be aware that soft tissue sarcomas in under 1s have a risk of consumptive coagulopathy (Kasabach Merrit syndrome). A full blood count is recommended in children under one with a suspected soft tissue sarcoma, especially if presenting with bruising and petechiae.

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved 81% (Delphi round 1)
- Rationale: While Kasabach Merrit phenomenon is an uncommon presentation, it is most common in CYP aged under-one year old. Therefore, the GDG and multidisciplinary team agreed it should be highlighted here.

Be aware of differences in skin presentations across skin tones

- Strength of evidence 4
- Recommendation form Conditional
- Consensus achieved Not applicable
- Rationale: The multidisciplinary team and GDG can recall this diagnostic pitfall being associated with delays to diagnosis. Therefore, the GDG and multidisciplinary workshop group agreed that this statement should be highlighted in the guideline. This statement is expert-initiated and was not sent out for consensus.

Requesting imaging for

(In primary care, requesting imaging should not delay referral/discussion with paediatrics)

In secondary care, request imaging for a CYP who has a persistent* or progressive lump/swelling with overlying skin changes.

- Strength of evidence 4
- Recommendation form Strong
- Consensus achieved 88% (Delphi round 1)

**persistent = present for two weeks or more*

4.8 Summary of recommendations for healthcare professionals

The Guideline Development Group (GDG) has also developed a one-page summary of guideline recommendations for healthcare professionals (see Appendix 1).

Implementation strategy and future work

5.1 Guideline implementation

The guideline implementation will be supported with the launch of the Child Cancer Smart awareness campaign (<https://www.cclg.org.uk/childcancersmart>) including clinical guidelines, quick reference and decision support tools for healthcare professionals.

All evidence generated by the Guideline Development Group (GDG) for the development of the guidelines, as well as any campaign outcomes, will be published and disseminated through professional conferences and in peer-reviewed journals.

5.2 Future review policy

The guideline is a stand-alone guideline written by the Guideline Development Group (GDG), jointly led by the CCLG and University of Nottingham. To ensure it provides high quality evidence to healthcare professionals across the country, this guideline requires five-yearly review. If new evidence becomes available or changes in referral pathways occur, then a full revision will be conducted in line with AGREE II criteria.

References

1. Ward ZJ, Yeh JM, Bhakta N, et al. Estimating the total incidence of global childhood cancer: a simulation-based analysis. *The Lancet Oncology* 2019;20(4):483-93. doi: [https://doi.org/10.1016/S1470-2045\(18\)30909-4](https://doi.org/10.1016/S1470-2045(18)30909-4)
2. Wang M, Bi Y, Fan Y, et al. Global incidence of childhood cancer by subtype in 2022: a population-based registry study. *eClinicalMedicine* 2025;89 doi: [10.1016/j.eclinm.2025.103571](https://doi.org/10.1016/j.eclinm.2025.103571)
3. Lethaby CD, Picton S, Kinsey SE, et al. A systematic review of time to diagnosis in children and young adults with cancer. *Arch Dis Child* 2013;98(5):349-55. doi: [10.1136/archdischild-2012-303034](https://doi.org/10.1136/archdischild-2012-303034) [published Online First: 20130309]
4. Shanmugavadivel D, Liu J-F, Ritzmann TA, et al. Quantifying diagnostic intervals and routes to diagnosis for children and young people with cancer in the UK (Childhood Cancer Diagnosis study, CCD): a population-based observational study. *The Lancet Regional Health – Europe* 2025;54 doi: [10.1016/j.lanepe.2025.101329](https://doi.org/10.1016/j.lanepe.2025.101329)
5. NICE. NICE guidance NG12: Suspected cancer: recognition and referral guidance for suspected cancer in children and young people. 2015. <https://www.nice.org.uk/guidance/ng12> (accessed 7/1/2026).
6. CCLG Referral guidance for suspected cancer in children and young people - A supporting resource for NICE guideline NG12 Suspected cancer: recognition and referral [Available from: <https://www.cclg.org.uk/Referral-guidance>.
7. NICE. National Institute for Health and Care Excellence. Suspected cancer: recognition and referral. [NICE guideline NG12] 2020
8. Scottish Intercollegiate Guidelines Network (SIGN). A guideline developer's handbook. Edinburgh: SIGN; 2019. (SIGN publication no. 50). [November 2019] [Available from: <https://www.sign.ac.uk/our-guidelines/sign-50-a-guideline-developers-handbook/>.
9. Ni Cheallaigh L, Liu JF, Fern L, et al. Clinical presentation of childhood soft tissue sarcomas: a systematic review and meta-analysis. *Arch Dis Child* 2024;109(2):113-20. doi: [10.1136/archdischild-2023-325875](https://doi.org/10.1136/archdischild-2023-325875) [published Online First: 20240122]
10. Ni Cheallaigh L, Ritzmann TA, Liu J-F, et al. Assessing and investigating children with suspected soft tissue sarcomas: an e-Delphi consensus process. *Archives of Disease in Childhood* 2026;archdischild-2026-330398. doi: [10.1136/archdischild-2026-330398](https://doi.org/10.1136/archdischild-2026-330398)
11. Abdel Wahab O, Qassemmyar A, Maillet M, et al. [Dermatofibrosarcoma protuberans in children]. *Ann Chir Plast Esthet* 2012;57(2):140-6. doi: <https://dx.doi.org/10.1016/j.anplas.2011.11.002>
12. An HY, Hong KT, Kang HJ, et al. Malignant peripheral nerve sheath tumor in children: A single-institute retrospective analysis. *Pediatric Hematology and Oncology* 2017;34(8):468-77. doi: [10.1080/08880018.2017.1408730](https://doi.org/10.1080/08880018.2017.1408730)
13. Benesch M, von Bueren AO, Dantonello T, et al. Primary intracranial soft tissue sarcoma in children and adolescents: a cooperative analysis of the European CWS and HIT study groups. *J Neurooncol* 2013;111(3):337-45. doi: <https://dx.doi.org/10.1007/s11060-012-1020-3>
14. Bots B, Eipel O, Terkovich L, et al. [Treatment results of pediatric soft tissue sarcomas at the 2nd Department of Pediatrics, Semmelweis University]. *Magy Onkol* 2018;62(4):222-29.

15. Bottcher A, Knopke S, Olze H, et al. Head and neck rhabdomyosarcoma in children: A 20-year retrospective study. *Laryngorhinootologie* 2018;97 (Supplement 2):S289-S90.
16. Boutroux H, Cellier C, Mosseri V, et al. Orbital rhabdomyosarcoma in children: a favorable primary suitable for a less-invasive treatment strategy. *J Pediatr Hematol Oncol* 2014;36(8):605-12. doi: <https://dx.doi.org/10.1097/MPH.0000000000000245>
17. Bravo-Ljubetic L, Peralta-Calvo J, Larranaga-Fragoso P, et al. Clinical Management of Orbital Rhabdomyosarcoma in a Referral Center in Spain. *J Pediatr Ophthalmol Strabismus* 2016;53(2):119-26. doi: <https://dx.doi.org/10.3928/01913913-20160122-02>
18. Cai MX, Pan C, Ye QD, et al. Clinical analysis of 51 cases with rare childhood soft tissue sarcomas. [Chinese]. *Zhonghua er ke za zhi = Chinese journal of pediatrics* 2016;54(12):917-22.
19. Cox CM, El-Mallawany NK, Kabue M, et al. Clinical characteristics and outcomes of HIV-infected children diagnosed with Kaposi sarcoma in Malawi and Botswana. *Pediatric Blood & Cancer* 2013;60(8):1274-80. doi: <https://dx.doi.org/10.1002/pbc.24516>
20. Croteau SE, Liang MG, Kozakewich HP, et al. Kaposiform hemangioendothelioma: atypical features and risks of Kasabach-Merritt phenomenon in 107 referrals. *J Pediatr* 2013;162(1):142-7. doi: <https://dx.doi.org/10.1016/j.jpeds.2012.06.044>
21. de Carvalho WRS, de Souza LL, Pontes FSC, et al. A multicenter study of oral sarcomas in Brazil. *Oral Dis* 2020;26(1):43-52. doi: <https://dx.doi.org/10.1111/odi.13211>
22. Dehner LP, Jarzembowski JA, Hill DA. Embryonal rhabdomyosarcoma of the uterine cervix: a report of 14 cases and a discussion of its unusual clinicopathological associations. *Mod Pathol* 2012;25(4):602-14. doi: <https://dx.doi.org/10.1038/modpathol.2011.185>
23. Demir HAK, T.; Ceyhan, M.; Yağcı-Küpeli, B.; Akyüz, C.; Cengiz, B.; Varan, A.; Kara, A.; Yalçın, B.; Seçmeer, G.; et al.,. Comparison of sulbactam-cefoperazone with carbapenems as empirical monotherapy for febrile neutropenic children with lymphoma and solid tumors. *Pediatric hematology and oncology* 2011;28(4):299-310. doi: 10.3109/08880018.2011.552937
24. Diaconescu S, Burlea M, Miron I, et al. Childhood rhabdomyosarcoma. Anatomico-clinical and therapeutic study on 25 cases. Surgical implications. *Rom J Morphol Embryol* 2013;54(3):531-7.
25. Dombrowski ND, Wolter NE, Robson CD, et al. Role of Surgery in Rhabdomyosarcoma of the Head and Neck in Children. *Laryngoscope* 2020;20:20. doi: <https://dx.doi.org/10.1002/lary.28785>
26. El-Mallawany NK, Kamiyango W, Slone JS, et al. Clinical Factors Associated with Long-Term Complete Remission versus Poor Response to Chemotherapy in HIV-Infected Children and Adolescents with Kaposi Sarcoma Receiving Bleomycin and Vincristine: A Retrospective Observational Study. *PLoS ONE [Electronic Resource]* 2016;11(4):e0153335. doi: <https://dx.doi.org/10.1371/journal.pone.0153335>
27. El-Nadi E, Elzomor H, Labib RM, et al. Childhood orbital rhabdomyosarcoma: Report from children's cancer hospital-57357-Egypt. *Journal of Solid Tumors* 2015;5(2):94-104.
28. Fasina O. Pattern of presentation and outcome of ophthalmic rhabdomyosarcoma in Ibadan. *Afr J Med Med Sci* 2013;42(2):165-9.
29. Fernandez-Pineda I, Spunt SL, Parida L, et al. Vaginal tumors in childhood: the experience of St. Jude Children's Research Hospital. *Journal of Pediatric Surgery* 2011;46(11):2071-5. doi: <https://dx.doi.org/10.1016/j.jpedsurg.2011.05.003>
30. Ferrari A, Miceli R, Casanova M, et al. The symptom interval in children and adolescents with soft tissue sarcomas. *Cancer* 2010;116(1):177-83. doi: <https://dx.doi.org/10.1002/cncr.24695>

31. Fortunati D, Kaplan J, Ponzzone A, et al. Desmoid Type Fibromatosis (DF): A 15-Years Experience at a Single Pediatric Institution in Argentina. *Pediatric blood & cancer* 2018;65
32. Giuseppucci C, Reusmann A, Giubergia V, et al. Primary lung tumors in children: 24 years of experience at a referral center. *Pediatr Surg Int* 2016;32(5):451-57.
33. Hausler SM, Stromberger C, Olze H, et al. Head and neck rhabdomyosarcoma in children: a 20-year retrospective study at a tertiary referral center. *J Cancer Res Clin Oncol* 2018;144(2):371-79. doi: <https://dx.doi.org/10.1007/s00432-017-2544-x>
34. Hemida R, Goda H, Abdel-Hady el S, et al. Embryonal rhabdomyosarcoma of the female genital tract: 5 years' experience. *J Exp Ther Oncol* 2012;10(2):135-7.
35. Hessissen L, Kanouni L, Kili A, et al. Pediatric rhabdomyosarcoma in Morocco. *Pediatric Blood & Cancer* 2010;54(1):25-8. doi: <https://dx.doi.org/10.1002/pbc.22173>
36. Hu PA, Zhou ZR. Clinical and imaging features of Kaposiform Hemangioendothelioma. *Br J Radiol* 2018;91(1086):20170798. doi: <https://dx.doi.org/10.1259/bjr.20170798>
37. Hu J, Wang H, Chang X, et al. Treatment for Abdominal Inflammatory Myofibroblastic Tumor in Children: An Analysis of Twelve Years of Experience. *Pediatric blood & cancer* 2018;65
38. Ibikunle AA, Taiwo AO, Braimah RO, et al. Orofacial Rhabdomyosarcoma: A 5-year Clinicopathologic Study from Sub-Saharan Africa. *Clinical Cancer Investigation Journal* 2018;7(2):56-61.
39. Iqbal CW, St Peter S, Ishitani MB. Pediatric dermatofibrosarcoma protuberans: multi-institutional outcomes. *J Surg Res* 2011;170(1):69-72. doi: <https://dx.doi.org/10.1016/j.jss.2011.01.042>
40. Ji Y, Yang K, Peng S, et al. Kaposiform haemangioendothelioma: clinical features, complications and risk factors for Kasabach-Merritt phenomenon. *Br J Dermatol* 2018;179(2):457-63. doi: <https://dx.doi.org/10.1111/bjd.16601>
41. Kadhim M, Oyoun NA, Womer RB, et al. Clinical and radiographic presentation of pelvic sarcoma in children. *Sicot-J* 2018;4:44. doi: <https://dx.doi.org/10.1051/sicotj/2018040>
42. Karamercan S, Okur A, Karadeniz C, et al. Non-rhabdomyosarcoma soft-tissue sarcomas in children: Experience from a single centre. *Pediatric Blood and Cancer* 2018;65 (Supplement 2):S327.
43. Koo MM, Lyratzopoulos G, Herbert A, et al. Association of Self-reported Presenting Symptoms With Timeliness of Help-Seeking Among Adolescents and Young Adults With Cancer in the BRIGHTLIGHT Study. *JAMA netw* 2020;3(9):e2015437. doi: <https://dx.doi.org/10.1001/jamanetworkopen.2020.15437>
44. Lim D, Hammond P, Hajivassiliou C, et al. Urinary retention in children: An unusual presentation which may herald a bladder tumour-a 20-year review. *Pediatric Blood and Cancer* 2015;62 (Supplement 4):S238-S39.
45. Livio V, Rose A, Zubizarreta P, et al. Pediatric orbital rhabdomyosarcoma: 20-year experience in a single institution in Argentina following the guidelines of siop trials. *Pediatric Blood and Cancer* 2013;60:93.
46. Maher OM, Khatua S, Mukherjee D, et al. Primary intracranial soft tissue sarcomas in children, adolescents, and young adults: single institution experience and review of the literature. *J Neurooncol* 2016;127(1):155-63. doi: <https://dx.doi.org/10.1007/s11060-015-2027-3>
47. Mathey MD, Peruzzo L, Cabero N, et al. Soft tissue sarcomas (STS) in the first year of life. A 15-year experience at a single pediatric institution in argentina. *Pediatric Blood and Cancer* 2018;65 (Supplement 2):S330.
48. Missaoui N, Landolsi H, Jaidene L, et al. Pediatric rhabdomyosarcomas in Tunisia. *Asian Pac J Cancer Prev* 2010;11(5):1325-7.

49. Okumu SB, Chindia ML, Gathece LW, et al. Clinical features and types of paediatric orofacial malignant neoplasms at two hospitals in Nairobi, Kenya. *J Craniomaxillofac Surg* 2012;40(1):e8-14. doi: <https://dx.doi.org/10.1016/j.jcms.2011.01.020>
50. Papillard-Marechal S, Brisse HJ, Pannier S, et al. [Pseudotumoral soft tissue masses in children and adolescents]. *Arch Pediatr* 2015;22(1):14-23. doi: <https://dx.doi.org/10.1016/j.arcped.2014.10.007>
51. Parida L, Fernandez-Pineda I, Uffman JK, et al. Clinical management of infantile fibrosarcoma: a retrospective single-institution review. *Pediatr Surg Int* 2013;29(7):703-8. doi: <https://dx.doi.org/10.1007/s00383-013-3326-4>
52. Perruccio K, Cecinati V, Scagnellato A, et al. Biliary tract rhabdomyosarcoma: a report from the Soft Tissue Sarcoma Committee of the Associazione Italiana Ematologia Oncologia Pediatrica. *Tumori* 2018;104(3):232-37. doi: <https://dx.doi.org/10.5301/tj.5000692>
53. Pinheiro M, Maia Ferreira A, Pinto A. Paediatric soft tissue sarcoma: A ten year review. *Arch Dis Child* 2014;99:A301.
54. Posso-De Los Rios CJ, Lara-Corrales I, Ho N. Dermatofibrosarcoma protuberans in pediatric patients: a report of 17 cases. *J Cutan Med Surg* 2014;18(3):180-5.
55. Reilly BK, Kim A, Pena MT, et al. Rhabdomyosarcoma of the head and neck in children: review and update. *Int J Pediatr Otorhinolaryngol* 2015;79(9):1477-83. doi: <https://dx.doi.org/10.1016/j.ijporl.2015.06.032>
56. Rifi H, Elboueiri A, Fehri R, et al. Child rhabdomyosarcoma: About Salah Azaiez institute patients. *Pediatric Blood and Cancer* 2011;57 (5):876.
57. Ronghe M, Sastry J, Murphy D, et al. West of scotland experience of rhabdomyosarcoma in children over a decade. *Pediatric Blood and Cancer* 2011;57 (5):793.
58. Sachedina A, Chan K, MacGregor D, et al. More than Grapes and Bleeding: An Updated Look at Pelvic Rhabdomyosarcoma in Young Women. *J Pediatr Adolesc Gynecol* 2018;31(5):522-25. doi: <https://dx.doi.org/10.1016/j.jpag.2018.01.006>
59. Shi J, Du J, Wu W, et al. [Clinical and imaging features of abdominal rhabdomyosarcoma of non-organ origin in children]. *Chung Hua Chung Liu Tsa Chih* 2016;38(11):845-51. doi: <https://dx.doi.org/10.3760/cma.j.issn.0253-3766.2016.11.009>
60. Siwillis EM, Dharse NJ, Scanlan T, et al. Pediatric Soft Tissue and Bone Sarcomas in Tanzania: Epidemiology and Clinical Features. *J* 2019;5:1-6. doi: <https://dx.doi.org/10.1200/JGO.18.00258>
61. Stefan DC, Stones DK, Wainwright L, et al. Kaposi sarcoma in South African children. *Pediatric Blood & Cancer* 2011;56(3):392-6. doi: <https://dx.doi.org/10.1002/pbc.22903>
62. Tsai YJ, Lin PY, Chew KY, et al. Dermatofibrosarcoma protuberans in children and adolescents: Clinical presentation, histology, treatment, and review of the literature. *J Plast Reconstr Aesthet Surg* 2014;67(9):1222-9. doi: <https://dx.doi.org/10.1016/j.bjps.2014.05.031>
63. Valdivielso-Ramos M, Torrelo A, Campos M, et al. Pediatric dermatofibrosarcoma protuberans in Madrid, Spain: multi-institutional outcomes. *Pediatr Dermatol* 2014;31(6):676-82. doi: <https://dx.doi.org/10.1111/pde.12371>
64. Varan A, Sen H, Aydin B, et al. Neurofibromatosis type 1 and malignancy in childhood. *Clin Genet* 2016;89(3):341-5. doi: <https://dx.doi.org/10.1111/cge.12625>
65. Wang HW, Qin XJ, Yang WJ, et al. Alveolar soft part sarcoma of the oral and maxillofacial region: clinical analysis in a series of 18 patients. *Oral Surg Oral Med Oral Pathol Oral Radiol* 2015;119(4):396-401. doi: <https://dx.doi.org/10.1016/j.oooo.2014.12.013>

66. Zhang WL, Zhang Y, Huang DS, et al. Clinical features of 39 children with head and neck rhabdomyosarcoma in a single medical center, and treatment outcomes. [Chinese]. Chinese Journal of Contemporary Pediatrics 2012;14(11):847-51.
67. Zhao M, Feng C, Wang JW, et al. Childhood rhabdomyosarcoma: A retrospective review of 23 cases. [Chinese]. Chinese Journal of Contemporary Pediatrics 2011;13(8):657-60.
68. Zorzi AP, Grant R, Gupta AA, et al. Cranial nerve palsies in childhood parameningeal rhabdomyosarcoma. Pediatric Blood & Cancer 2012;59(7):1211-4. doi: <https://dx.doi.org/10.1002/pbc.24128>
69. Deyrup AT, Miettinen M, North PE, et al. Pediatric cutaneous angiosarcomas: a clinicopathologic study of 10 cases. Am J Surg Pathol 2011;35(1):70-5. doi: <https://dx.doi.org/10.1097/PAS.0b013e3181ffd9d5>
70. Report of the Independent Cancer Taskforce. Achieving World-Class Cancer Outcomes: A Strategy for England 2015-2020 [Available from: http://www.cancerresearchuk.org/sites/default/files/achieving_world-class_cancer_outcomes_-_a_strategy_for_england_2015-2020.pdf
71. Department of Health. Referral guidelines for suspected cancer. HSC 2000/013. London: Department of Health; 2000.
72. Galloway P, Arumugam J, Hargreaves J, et al. How effective is the 2 week urgent referral pathway in identifying paediatric cancer? experience from a general paediatric hospital and literature review. Archives of Disease in Childhood 2016;101(Suppl 1):A8. doi: 10.1136/archdischild-2016-310863.13
73. Mant JE, Nanduri V, Francis D. The 2 week urgent referral pathway for suspected cancer: the paediatric experience in a local oncology shared-care centre. Archives of Disease in Childhood 2011;96(Suppl 1):A85. doi: 10.1136/adc.2011.212563.198
74. Würtemberger J, Ripperger T, Vokuhl C, et al. Genetic susceptibility in children, adolescents, and young adults diagnosed with soft-tissue sarcomas. European Journal Medical Genetics. 2023;66(5):104718. doi: 10.1016/j.ejmg.2023.104718.

Appendix 1. Summary of recommendation

Diagnosis of soft tissue sarcomas in children and young people

A summary for healthcare professionals

Consider a soft tissue sarcoma in any child or young person presenting with:

- Unexplained lump
- Swelling
- Pain
- Skin changes over the lump/swelling

Associated symptoms:

- Limbs: lump, limp, restricted movement or altered function
- Head and neck: localised eye swelling, cranial nerve deficit, impaired vision, ear swelling or pain
- Abdomen and pelvis: urinary symptoms, abdominal /flank distension, genital lump/swelling, vaginal bleeding, change in bowel habit, obstructive jaundice
- Systemic symptoms: weight loss, fever, malaise
- Bruising/bleeding, especially in CYP aged under one year
- Back pain (neck to sacrum) with or without signs of spinal cord compression

Ask about predisposing factors

- Neurofibromatosis 1
- Li-Fraumeni syndrome
- DICER1
- Family history of breast or ovarian cancer at a young age
- A sibling with childhood cancer

Assess these children with:

History:

- Associated symptoms
- Any predisposing factors
- Family history

Examination of:

- Lump and regional lymph node
- Skin (birth marks, e.g. café-au-lait spots)
- Neurological examination
- Age-appropriate weight and growth

Assessment pitfalls

- Symptoms are usually progressive and can cause other seemingly unrelated symptoms due to pressure on other structures, such as respiratory distress, spinal cord compression or bowel obstruction.
- In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.

Referral from primary care

- Discuss all suspected cases with paediatrics via telephone to advise best route to be seen.

Imaging:

- Be aware that ultrasound imaging is not always recommended for a soft tissue sarcoma.
- In secondary care, discuss with paediatric radiologist to decide most suitable imaging.

Unexplained lump

- Soft tissue sarcomas can occur anywhere in or on the body.
- They tend to be fixed and hard on palpation.
- They can be associated with pain, but pain is not always present.
- Sometimes they can have overlying changes in the skin.
- Ask about the presence of the other symptoms of cancer: weight loss, fever, weight loss, back pain, bowel/bladder/renal symptoms.
- CYP presenting with an unexplained lump.
- Depending on where the lump is, ask about the other symptoms of cancer: head and neck symptoms (localised eye swelling, cranial nerve palsies, hearing loss), respiratory symptoms (new onset cough, haemoptysis) and abdomen/pelvis symptoms (urinary symptoms, constipation, weight loss).

Imaging required for:

- Unexplained persistent* and growing lump
- Unexplained lump with another symptom from the

**persistent = present for two weeks or more*
(In primary care, requesting an ultrasound should not

Diagnostic pitfalls:

- Attributing lump to an injury without a clear history
- Attributing lumps to lipomas or haemangiomas with uncertainty and no improvement, always discuss second opinion or review.
- Misdiagnosis of a benign mass on imaging, lead reported by paediatric radiologists, and any diagnosis should be discussed at the appropriate paediatric

Pain

- In soft tissue sarcomas, pain is often associated
- Depending on where the lump or swelling is, pain
- Be aware of referred pain.
- Be aware that children aged younger than 4 years are frequently unable to describe or localise pain. Weight bearing, may indicate bone pain. Look for
- Ask and examine for the other signs and symptoms: skin changes, birth marks (café au lait) fever, weight dysfunction if back pain is present) in CYP with pain

Imaging required for:

- A painful, persistent* or progressive lump or swelling
- *persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not

Diagnostic pitfalls:

- Not performing a full examination for those pres

on the body and present as a lump.

t always present.
the skin.

of a soft tissue sarcoma (swelling, pain, skin changes,
ctile dysfunction, birth marks e.g café au lait spots) in a

presence of other associated symptoms including head
nial nerve deficit, impaired vision, ear pain and swelling
et of swallowing difficulties / choking, stridor or snoring
ms, abdominal distension, genital mass, vaginal bleeding).

ne symptom checklist.

delay referral/discussion with paediatrics)

ory
without typical appearance. If there is diagnostic
s with a paediatrician or a paediatric surgeon for a

ing to an incorrect surgical procedure. Imaging should be
gnostic uncertainty
ic oncology MDT prior to surgery.

with the lump or swelling.
n can be neuropathic.

rs, or those with communication difficulties,
n; their behaviour e.g. withdrawal, holding their leg, not
r these signs on examination.
ms suggestive of a soft tissue sarcoma (lump, swelling,
ght loss, back pain and bowel/bladder/erectile
pain.

swelling.

delay referral/discussion with paediatrics)

enting with pain that may be referred.

Swelling

- Swelling in soft tissue sarcoma can occur on any part of the body.
- Ask about the presence of the other symptoms of a soft tissue sarcoma (lump, pain, skin changes, birth marks (café au lait spots) fever, weight loss, back pain and bowel/bladder/erectile dysfunction).
- Ask about the presence of head and neck symptoms (localised eye swelling, cranial nerve deficit, impaired vision, ear pain or swelling or localised hearing loss) if the lump is in the head/neck area.

Imaging required for:

- Any persistent swelling* rapidly increasing in size, including an eye or ear swelling.
- Any persistent swelling* not resolving despite treatment with regular anti-inflammatories OR antibiotics.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.)

Diagnostic pitfalls:

- Attributing a red warm swelling to infection despite no improvement with antibiotics. Arrange to see the CYP at the end of the course of antibiotics to assess response. If there has been no response, consider discussion with secondary care for advice or referral.

Skin changes over the lump/swelling

- Skin changes can include erythema, thinning of the overlying skin, ulceration, hyperpigmentation, fungating lesion.
- In under-ones, there is risk of consumptive coagulopathy which can cause petechiae, bruising and bleeding.
- Be aware that these skin changes can look different across different skin tones.
- Ask about the presence of the other symptoms of a soft tissue sarcoma in a CYP presenting with skin changes over a lump or swelling pain (fever, weight loss, back pain and bowel/bladder/erectile dysfunction if back pain is present).

Imaging required for:

- A CYP who has a persisting * or progressive lump/swelling with overlying skin changes.

**persistent = present for two weeks or more*

(In primary care, requesting an ultrasound should not delay referral/discussion with paediatrics.)

Diagnostic pitfalls:

- Be aware that soft tissue sarcomas in under 1s have a risk of consumptive coagulopathy (Kasabach-Merritt syndrome). A full blood count is recommended in children under one with a suspected soft tissue sarcoma, especially if presenting with bruising and petechiae.
- Be aware of differences in skin presentations across skin tones.

Appendix 2.

GDG members, multidisciplinary workshop and Delphi participants

GDG Members

1. Dr Lorna Ni Cheallaigh (Great Ormond Street Institute of Child Health, University College London, London, UK)
2. Ms Jo-Fen Liu (Early Diagnosis Project Manager and Statistician, CCLG: The Children & Young People's Cancer Association, Leicester, UK)
3. Dr Shaarna Shanmugavadivel (CCLG Early Diagnosis Fellow, Lifespan and Population Health, University of Nottingham, Nottingham, UK)
4. Dr Tim Ritzmann (Clinical Associate Professor in Paediatric Neuro-Oncology and Molecular Biology and Honorary Consultant Paediatric Oncologist, Consultant Paediatric Oncologist, Children's Brain Tumour Research Centre, University of Nottingham, Nottingham, UK)
5. Emeritus Professor David Walker (Children's Brain Tumour Research Centre, University of Nottingham, Nottingham, UK)

Multidisciplinary Workshop Participants

1. Dr Damian Wood (Consultant Paediatrician, Nottingham University Hospitals NHS Trust).
2. Dr Jill Sussens (Consultant Paediatrician, Nottingham University Hospitals NHS Trust).
3. Dr Paul Nathan (GP, Hollybrook Medical Centre, Derby).
4. Dr Hugh Bishop (Consultant Paediatrician Oncologist, Royal Aberdeen Children's Hospital)
5. Emeritus Prof David Walker (Children's Brain Tumour Research Centre, University of Nottingham, Nottingham, UK)
6. Dr Tim Ritzmann (Clinical Associate Professor in Paediatric Neuro-Oncology & Molecular Biology and Honorary Consultant Paediatric Oncologist Children's Brain Tumour Research Centre School of Medicine, University of Nottingham, Nottingham UK)
7. Dr Lynda Walton (Consultant in Paediatric Emergency Medicine, Nottingham University Hospitals NHS Trust)
8. Dr Graham Johnson (Consultant in Paediatric Emergency Medicine, University Hospitals of Derby and Burton NHS Foundation Trust)
9. Mr Tristan Boam (Specialist Registrar CCT Paediatric Surgery, Nottingham University Hospitals NHS Trust)
10. Mr Ashley Ball-Gamble (CEO, CCLG: The Children & Young People's Cancer Association, Leicester, UK)
11. Dr Shaarna Shanmugavadivel (CCLG Early Diagnosis Fellow, Lifespan and Population Health, University of Nottingham, Nottingham, UK)
12. Dr Lorna Ni Cheallaigh (Great Ormond Street Institute of Child Health, University College London, London, UK)
13. Ms Jo-Fen Liu (Early Diagnosis Project Manager, CCLG: The Children & Young People's Cancer Association, Leicester, UK)
14. Mrs Sue Franklin (Parent)
15. Mr Neil Ranasinghe (Parent)
16. Dr Suzi Tugnait (University College of London)

Child Cancer Smart Oncology Research Lead

Dr Tim Ritzmann, Clinical Associate Professor in Paediatric Neuro-Oncology & Molecular Biology and Honorary Consultant Paediatric Oncologist, Children's Brain Tumour Research Centre, University of Nottingham, Nottingham, UK

Delphi Consensus Participants

General Paediatricians

- | | | |
|-------------------------|----------------------------|-----------------------------|
| 1. Dr Anand Srivastava | 9. Dr Bindu Koodiyedath | 17. Dr Katherine Piccinelli |
| 2. Dr Jill Sussens | 10. Dr Caroline Brown | 18. Dr Kim Neuling |
| 3. Dr Rebecca Proudfoot | 11. Dr Caroline Scott-Lang | 19. Dr Louise Wells |
| 4. Dr Ajanta Kamal | 12. Dr Elinor Sefi | 20. Dr Marie-Louise Millard |
| 5. Dr Alison Davies | 13. Dr Eva Tsouana | 21. Dr Salma Ali |
| 6. Dr Angela Gilbert | 14. Dr Gemma Williams | 22. Dr Sarah Thompson |
| 7. Dr Arindam Das | 15. Dr Hannah Baynes | 23. Dr Tasnim Arif |
| 8. Dr Beth Cheesebrough | 16. Dr Jane Hoddes | |

Paediatric Emergency Medicine

- | | | |
|-----------------------|-----------------------|------------------------|
| 1. Dr Clare Dieppe | 5. Dr Helen Milne | 9. Dr Simon Li |
| 2. Dr Colin Gilhooley | 6. Dr Jane Bayreuther | 10. Dr Vikram Baicher |
| 3. Dr Dani Hall | 7. Dr Scott Hendry | 11. Dr Michaela Lazner |
| 4. Dr David Smith | 8. Dr Segn Nedd | |

Radiologists

- | | | |
|--------------------|------------------------|---------------------|
| 1. Dr Angela Byrne | 3. Dr Matt Smedley | 5. Dr Sara Harrison |
| 2. Dr Geoff Hide | 4. Dr Rachael Harrison | 6. Dr Saurabh Singh |

Paediatric Surgeons

- | | | |
|--------------------------------|-------------------------|-------------------------|
| 1. Miss Bhanu Lakshminarayanan | 4. Mr Craig Gerrand | 7. Mr Ross Craigie |
| 2. Mr Richard Hewitt | 5. Mr Max Pachi | 8. Miss Sarah Braungart |
| 3. Mr Basil Bekdash | 6. Mr Michael Jacovides | 9. Mr Timothy Rogers |

GPs

- | | | |
|-------------------------|-----------------------|---------------------------|
| 1. Dr Ben Milton | 4. Dr Katherine Teare | 7. Dr Penny Edrich |
| 2. Dr Claire Grimstvedt | 5. Dr Kathryn Bannell | 8. Dr Rebecca Chellaswamy |
| 3. Dr Edward Rendell | 6. Dr Lucy Follis | 9. Dr Sharon Healy |

Paediatric Oncologists

- | | | |
|-------------------------------|--------------------------|----------------------------|
| 1. Dr Amy Mitchell | 5. Dr Bernadette Brennan | 9. Dr Henry Mandeville |
| 2. Dr Quentin Campbell Hewson | 6. Dr Courtney Willis | 10. 10. Dr Madeleine Adams |
| 3. Dr Amos Burke | 7. Dr Gerard Millen | |
| 4. Dr Anna Jenkins | 8. Dr Helen Rees | |

Appendix 3.

Strength of evidence and recommendation

Levels of Evidence

Level	Description
1++	High-quality meta-analyses, systematic reviews of randomized controlled trials (RCTs), or RCTs with a very low risk of bias.
1+	Well-conducted meta-analyses, systematic reviews of RCTs, or RCTs with a low risk of bias.
1-	Meta-analyses, systematic reviews of RCTs, or RCTs with a high risk of bias.
2++	High-quality systematic reviews of case-control or cohort studies; high-quality case-control or cohort studies with a very low risk of confounding or bias and a high probability that the relationship is causal.
2+	Well-conducted case-control or cohort studies with a low risk of confounding or bias and a moderate probability that the relationship is causal.
2-	Case-control or cohort studies with a high risk of confounding or bias and a significant risk that the relationship is not causal.
3	Non-analytic studies, e.g., case reports, case series.
4	Expert opinion.

Grade of Recommendations

Grade	Description
A	At least one meta-analysis, systematic review, or RCT rated as 1++, and directly applicable to the target population; or a body of evidence consisting principally of studies rated as 1+, directly applicable to the target population, and demonstrating overall consistency of results.
B	A body of evidence including studies rated as 2++, directly applicable to the target population, and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 1++ or 1+.
C	A body of evidence including studies rated as 2+, directly applicable to the target population and demonstrating overall consistency of results; or extrapolated evidence from studies rated as 2++.
D	Evidence level 3 or 4; or extrapolated evidence from studies rated as 2+.

Forms of Recommendation (SIGN 50)^[8]

Judgement	Recommendation
Undesirable consequences clearly outweigh desirable consequences	Strong recommendation against
Undesirable consequences probably outweigh desirable consequences	Conditional recommendation against
Desirable consequences probably outweigh undesirable consequences	Conditional recommendation for
Desirable consequences clearly outweigh undesirable consequences	Strong recommendation for

‘Strong’ recommendations should be made where there is confidence that, for the vast majority of people, the intervention/action will do more good than harm (or more harm than good). The recommendation should be clearly directive and include **‘should/ should not’** in the wording.

‘Conditional’ recommendations should be made where the intervention/action will do more good than harm for most patients but may include caveats e.g. on the quality or size of the evidence base, or patient preferences. Conditional recommendations should include **‘should be considered’** in the wording.

Notes



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Produced by the Child Cancer Smart Guideline Development Group.

We are CCLG: The Children & Young People's Cancer Association. We unite the children and young people's cancer community, driving collective action and progress. Powered by expertise, we work together to create a brighter future for children and young people with cancer.

Research is the key to better treatments, improved care, and potential cures. We fund and lead world-class research, fuelling groundbreaking work led by brilliant minds. Collaboration is at the heart of our approach - bringing together the right people and organisations to drive progress and deliver real impact.

We provide trusted information and guidance for children and young people with cancer, their families, and everyone supporting them. Our expertise helps them navigate the challenges of cancer and its impact, offering reassurance and clarity when it's needed most.

Through our professional membership, we bring together the brightest minds in childhood cancer, creating a national network that drives progress. Together, we shape better treatment and care - developing guidelines, sharing knowledge, offering expert advice, leading pioneering research, and creating essential resources and education for professionals. Our collective expertise sets the standard, advocating for excellence at every level - local, national, and global.

These guidelines are to inform and are for use at the sole discretion of treating healthcare professionals who retain professional responsibility for their actions and treatment decisions. Guidelines that are printed or stored in a local system are uncontrolled documents. Guidelines should be accessed from the CCLG website to ensure the latest version is used.