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Young People's  
Cancer Association



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# A guideline to assist healthcare professionals in the assessment of children and young people who may have an abdominal tumour

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# Guideline background, aim and scope

## Background

The Childhood Cancer Diagnosis Study studied time to diagnosis for children and young people across the UK. Abdominal tumours account for around 15% of childhood cancers and there is strong evidence that children in the UK with a Wilms tumour have a significantly larger volume, a more advanced tumour stage at diagnosis and poorer survival compared with their European counterparts<sup>[1]</sup>. This highlighted a need to urgently review the clinical guidance available to health care professionals.

The 2015 National Institute for Health and Care Excellence (NICE) published a guideline on ‘Suspected cancer: recognition and referral’ which covers all ages<sup>[2]</sup>. There is a real need for paediatric-specific guidance, as adult cancers manifest and present differently. This current 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 and a referral from primary to secondary care can add significant length to the patient’s diagnostic journey. Timely ultrasound imaging is essential to identify children who need further investigations for abdominal tumours and so guidance was required on indications for and appropriate waiting times to ultrasound 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 or haematologists. 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<sup>[3]</sup>. 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 similar to that produced for childhood brain tumours is needed to empower clinicians to make decisions about those who need investigation and accelerate referrals for children and young people with high-suspicion of abdominal tumour promoting earliest possible diagnosis.

## Aim of the guideline

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

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

## Clinical health questions

The guideline was devised to address the following questions:

1. What are the symptoms and signs that CYP with abdominal tumours present with?
2. Given that the initial symptoms and signs of an abdominal tumour may occur with other less serious childhood conditions, how can healthcare professionals distinguish those CYP who may have an abdominal tumour from the majority who do not?
3. What is the best way to clinically assess a CYP presenting with these symptoms and/or signs?
4. What symptoms and/or signs in CYP increase the likelihood of an abdominal tumour 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 symptoms and/or signs, what is an appropriate maximum waiting time to imaging?
7. Are there specific presentations of childhood abdominal tumours that are repeatedly associated with diagnostic difficulty?
8. Are there other barriers to diagnosis in childhood abdominal tumours and if so, how can these be addressed?

## Scope and target population

The guideline is applicable to all children and young people aged 0-18 years who present with symptoms and/or signs that could be due to an abdominal tumour 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, 2019)<sup>[4]</sup>.

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 abdominal tumour. 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 the 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 who 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 abdominal tumours:

- 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)

## Funding

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The systematic reviews were funded by Cancer Research UK Innovation Grant awarded to Dr Sharna Shanmugavadivel (C59357/A22874) and CCLG: The Children & Young People's Cancer Association.

## 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 and implementation process.

# Methods

## 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).

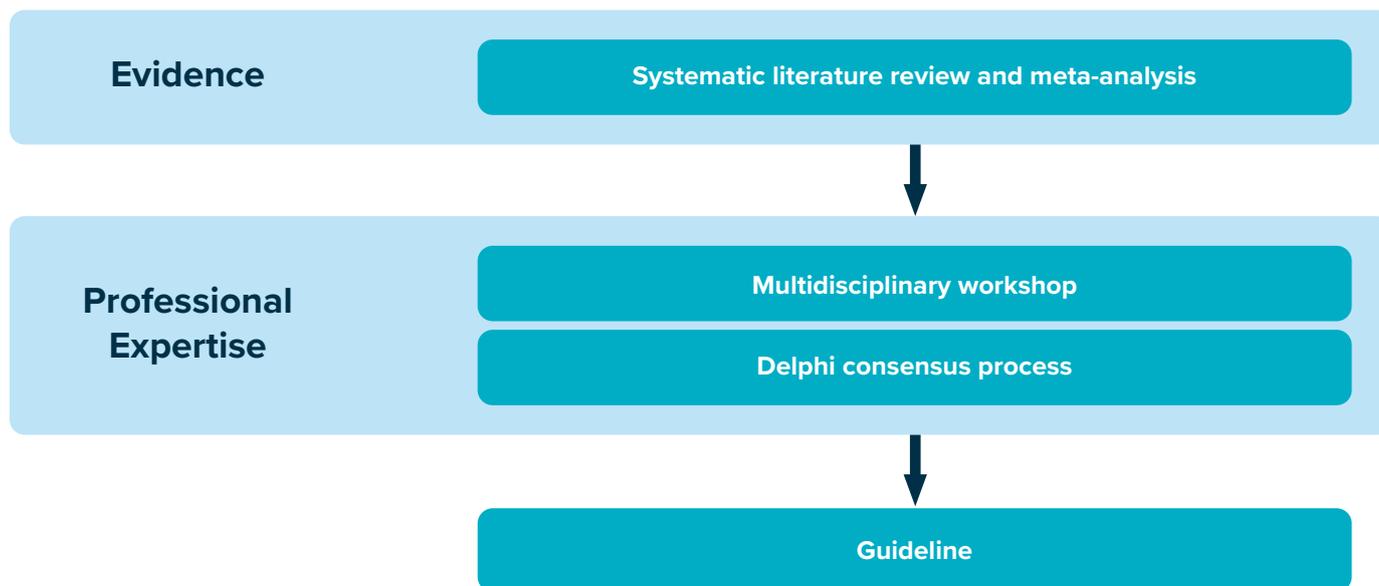


Figure 1. Guideline development methodology

The initial stage comprised a systematic review and meta-analysis to appraise current evidence on childhood abdominal tumour presentation. 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.

## Impact of the COVID-19 pandemic

The systematic review and meta-analysis were originally carried out in 2019<sup>[5]</sup>. The multi-disciplinary workshop and Delphi consensus survey was subsequently conducted in 2019 and early 2020, with the findings published in 2023<sup>[6]</sup>. However, the guideline development was delayed due to the COVID-19 pandemic. To ensure the inclusion of up-to-date evidence in this guideline, another search was carried out in 2023. The combined analysis, which provided contemporary information and evidence regarding the presentation, has since been published<sup>[7]</sup>. The updated search in 2023 and meta-analysis did not identify any new features but rather ensured the evidence used in the Delphi process was still up to date.

## Systematic review and meta-analysis

**Clinical Question: What are the symptoms and signs that children with abdominal tumours develop?**

A systematic review and meta-analysis of the presenting signs and symptoms of abdominal tumours 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<sup>[7]</sup>.

## 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 abdominal tumours and to advise on appropriate referral pathways, imaging indications and acceptable waiting times.

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 symptoms could be due to a abdominal tumour, i.e. their specificity?
- What factors influence the specificity of a sign and symptom?
- What are appropriate thresholds for referral and selection for ultrasound imaging for a child presenting with this sign or symptom?
- What would they regard as best practice for referral and imaging of a child presenting with this sign and 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.

## 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<sup>[6]</sup>.

**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: The Children & Young People's Cancer Association (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.

## Strength of evidence and recommendation

The levels of evidence and grades of recommendations (Appendix 5) are based on the SIGN 50: A Guideline Developer's Handbook<sup>(4)</sup>. Recommended best practice are based on the clinical experience of the GDG.

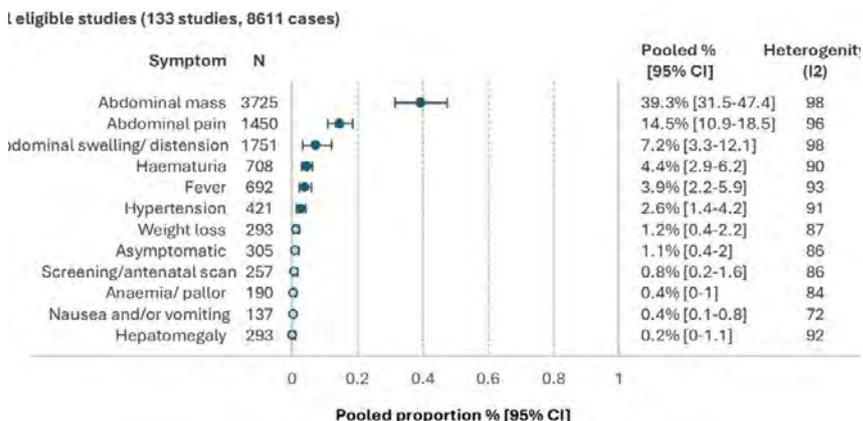
# The evidence

## Systematic review and meta-analysis

The initial search of MEDLINE and EMBASE identified 19,831 titles published between January 2005 and December 2023. After title and abstract screening, 669 were selected for full-text review and 133 studies included in the final analysis<sup>[8-140]</sup>. These studies provided data on 8611 cases of childhood abdominal tumours, across 42 different countries. The quality was comprehensively evaluated and summarised. A total of 147 symptoms/signs were recorded and those with pooled proportions 2% or more of the studied population are reported in Figures 2-4.

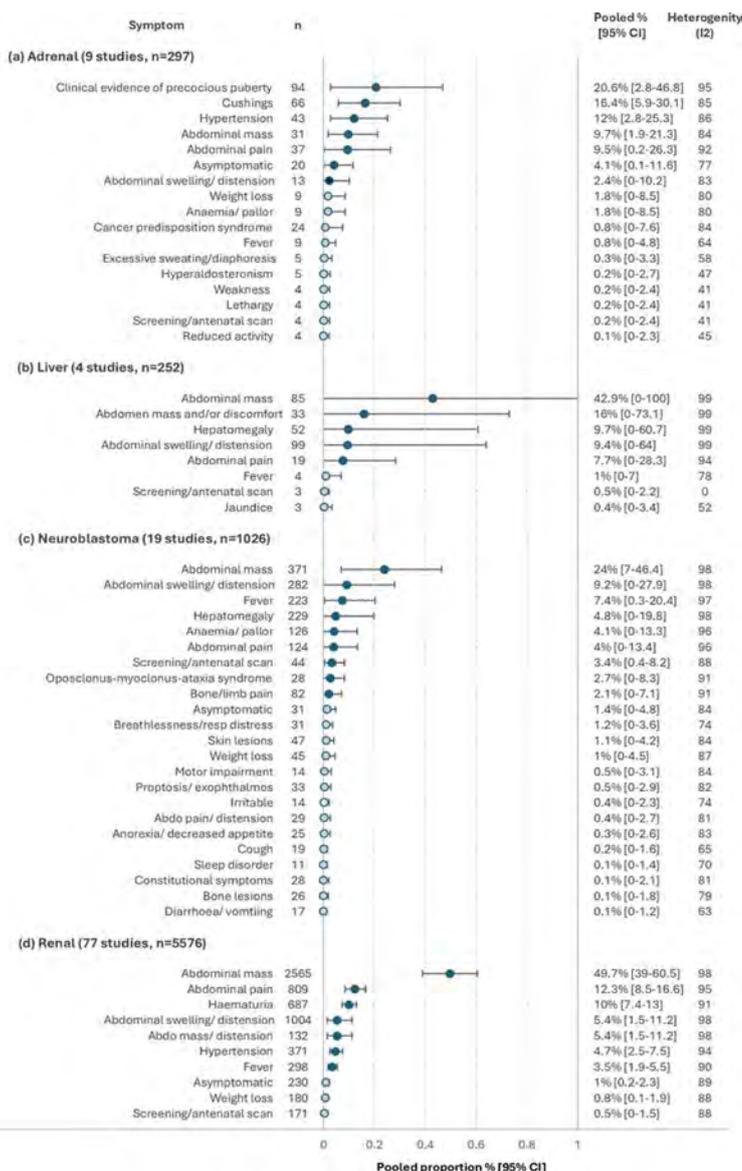
**Figure 2.** Pooled proportions for the most frequently reported pre-diagnostic symptoms/signs in the whole cohort.

Hollow circles represent symptoms/signs with a pooled proportion of <2%.



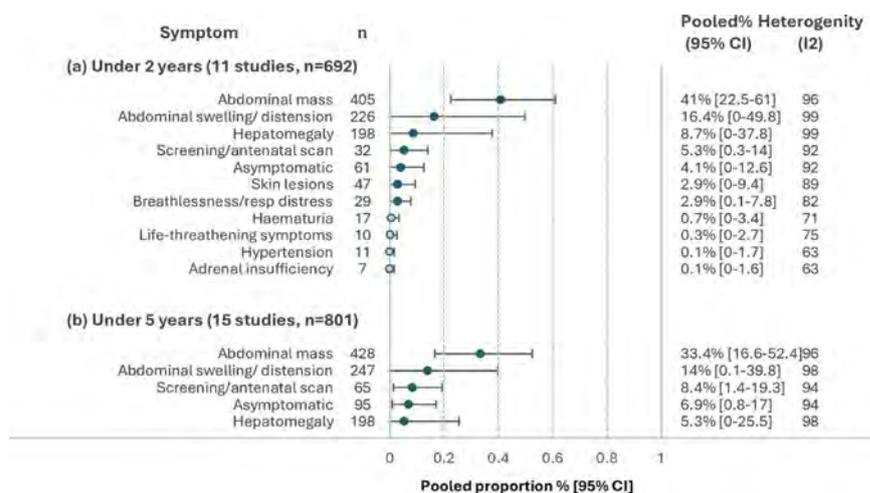
**Figure 3.** Pooled proportions for the most frequently reported pre-diagnostic symptoms/signs for abdominal tumours in (a) adrenal gland, (b) liver, (c) renal and (d) neuroblastoma.

Hollow circles represent symptoms/signs with a pooled proportion of <2%.



**Figure 4.** Pooled proportions for the most frequently reported pre-diagnostic symptoms/signs for abdominal tumours diagnosed in (a) under 2 years of age and (b) under 5 years of age.

Hollow circles represent symptoms/signs with pooled proportions of <2%.



## Core and associated symptoms

Abdominal tumours encompass a large number of different tumours, namely Wilms' tumour, neuroblastoma, hepatoblastoma, abdominal lymphomas, soft tissue sarcomas and germ cell tumours.

The following symptoms and signs are all associated with childhood abdominal tumours. Their presence should alert the clinician to this possibility:

### Core symptoms

- Abdominal mass
- Abdominal pain
- Haematuria
- Abdominal distension

### Associated symptoms

- Weight loss
- Pallor/petechiae
- Fever
- Malaise
- Jaundice
- Hepatosplenomegaly
- Bone pain
- Neurological symptoms from spinal cord compression
- Bladder/bowel/erectile dysfunction due to spinal cord compression
- Urinary retention

## Delphi consensus

The Delphi consensus process comprised of 37 statements describing best practice (referral, imaging, predisposing factors), the presenting features of childhood abdominal tumours (general, abdominal mass, abdominal pain, haematuria, abdominal distension), factors that could be used to discriminate these tumours from other less serious conditions and possible referral pathways for children with symptoms or signs suggestive of abdominal tumours.

The initial round achieved a consensus in 95% of the statements sent out for review and all statements reached consensus within two rounds; no statement was rejected. The full Delphi consensus process methodology and results for best practice and abdominal tumour statements have been published<sup>[6]</sup>.

**Table 1. Best practice statements and percentages consensus**

<b>General</b>	1. Explicitly ask young people, parents and carers about their concerns regarding what the cause of symptoms are in any consultation.	99.0%
	2. If a parent/carer expresses concerns about an abdominal tumour this should be reviewed carefully. If a tumour is unlikely, explain why and give appropriate safety netting advice.	72.0%
	3. Any healthcare professional deciding to review a patient to diagnose or exclude cancer should ensure that the timing of the review does not exceed the national 4-week limit to access a diagnostic test and obtain the result.	89.8%
	4. Offer a telephone or in person interpreter if the patient, parent / carer or healthcare professional are not fluent in English or Welsh.	95.9%
	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.	90.7%
	6. Be aware that the presence of complex neuro-disabilities or other communication difficulties (eg, 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.	98.0%
<b>Referral</b>	7. In primary care, discuss concerns with your local consultant paediatrician hotline or the paediatric consultant on call the same day if there is a high index of suspicion regarding a possible abdominal tumour in a CYP.	90.1%
	8. Discuss concerns over the telephone with the consultant paediatrician hotline or local equivalent service before referring a CYP from primary care in which differential diagnosis includes a possible tumour to ensure the CYP is seen within the most suitable timeframe (ideally within 2 weeks).	89.6%
	9. Abdominal tumours can progress rapidly over days, increasing in size and causing pressure effects (e.g. breathing difficulties or bowel obstruction and ischaemia). If there is suspicion of an abdominal tumour this should be discussed in person with a paediatrician to decide the most appropriate time for review, which will often be the same day.	92.7%
<b>Imaging</b>	10. Request an ultrasound of the abdomen and pelvis as the initial investigation for a CYP who has a suspected abdominal tumour. This should not delay referral from primary care.	80.4%
<b>Predisposing factors</b>	11. Be aware that some predisposing factors are associated with an increased risk of childhood abdominal tumours. Verify the presence of predisposing factors with parents/patients as they may lower the threshold for referral and investigation.	83.5%

\*CYP, children and young people.

**Table 2. Abdominal tumour statements and percentages consensus**

<b>General</b>	12. Take a detailed history including the presence of any of the other symptoms on the list above, any predisposing factors and a family history.	94.8%
	13. Examine the abdomen, including external genitalia and hernia orifices, visualise and palpate the spine and perform a neurological examination in a CYP with signs/symptoms that may be due to an abdominal tumour.	93.6%
	14. Perform a urine dipstick to exclude infection and identify any protein or blood that would warrant referral to secondary care.	89.7%
	15. Consider recording blood pressure if the correct size cuff is available to identify hypertension.	74.2%
	16. Be aware that weight loss can be a sign of an abdominal tumour. Measure weight and compare to any previous measurements in CYP with signs or symptoms suggestive of an abdominal tumour. Plot these measurements on age-appropriate growth charts if available to you to monitor change when reviewing symptoms.	88.3%
<b>Abdominal mass</b>	17. Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, haematuria, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with an abdominal mass.	99.0%
	18. 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 when a CYP presents with refusal to weight bear.	94.9%
	19. Be aware that diagnostic delay is associated with failure to perform an abdominal examination in a child who is distressed/crying. Offer to examine the child once they have settled or ask a colleague to perform the examination.	94.8%
	20. Request ultrasound imaging of the abdomen and pelvis for a CYP with a palpable abdominal mass (unless this is felt to be faeces, in which case a review of the CYP after disimpaction is important).	83.6%
	21. Request ultrasound imaging of the abdomen and pelvis for a CYP with suspected hepatomegaly or splenomegaly on examination.	83.5%
<b>Abdominal pain</b>	22. Ask about the presence of the other symptoms of an abdominal tumour (haematuria, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with persistent abdominal pain.	92.8%
	23. Examine the abdomen in a CYP with abdominal pain to elicit any masses or hepatomegaly and/or splenomegaly.	99.0%
	24. Be aware that if the tumour is retroperitoneal, the presenting complaint may be back pain. Examine the abdomen in a CYP presenting with back pain.	95.9%
	25. Be aware that delayed diagnosis has been associated with attributing abdominal pain to constipation despite no improvement with laxatives. Assess response to laxatives by reviewing the CYP at regular intervals and taking a full history and examining their abdomen.	87.7%

	26. Request ultrasound imaging for persistent abdominal pain with one or more other symptoms that may be due to an abdominal tumour (abdominal distension, mass, haematuria, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction).	86.6%
<b>Haematuria</b>	27. Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, mass, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with haematuria in the absence of another known cause.	96.9%
	28. Do a careful abdominal examination to elicit any potential retroperitoneal mass in all CYP with haematuria.	92.7%
	29. Be aware that delayed diagnosis has been associated with assuming persistent haematuria is due to a UTI despite no response to antibiotics. Arrange to see the CYP again at the end of the course of antibiotics to assess the response. If there is no response, consider discussion with paediatrician for advice.	82.4%
	30. Request ultrasound imaging for unexplained persistent (occurring for 2 weeks or more) macroscopic haematuria.	87.7%
	31. Request ultrasound imaging of the abdomen and pelvis for haematuria plus one or more other symptoms that may be due to an abdominal tumour (abdominal pain, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction).	87.6%
<b>Abdominal distension</b>	32. Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, haematuria, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with an abdominal distension.	97.9%
	33. Be aware that abdominal distension caused by a tumour will not fluctuate but will increase in size progressively over time. Arrange another appointment to review the symptoms and re-examine the CYP.	83.5%
	34. Be aware that delayed diagnosis has been associated with attributing abdominal distension to constipation despite no improvement with laxative treatment. Assess response to treatment in these CYP by seeing them at regular intervals.	84.5%
	35. Be aware that delayed diagnosis has been associated with failure to examine the abdomen at review of a CYP on treatment for constipation. Palpate the abdomen in CYP who are being reviewed for constipation.	94.9%
	36. Request ultrasound imaging of the abdomen and pelvis for a CYP with rapidly increasing abdominal distension. This should not delay referral from primary care.	83.5%

\*CYP, children and young people. UTI, urinary tract infection.

# Clinical Guideline

## Best practice – consultation

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

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	99% (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.

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**If a parent/carer expresses concerns about a possible abdominal 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	72% (Delphi Round 1)

### Rationale

Parents / carers of children with abdominal tumours 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.

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**Any healthcare professional deciding to review a patient to diagnose or exclude cancer should ensure that the timing of the 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	89.8% (Delphi Round 1)

### Rationale

Symptom progression occurs with childhood cancers, therefore early review is recommended to facilitate detection of any additional 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<sup>[141]</sup>.

**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	95.9% (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 / carer 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	90.7% (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 abdominal tumours. 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	98.0% (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.

## Best practice – referral

**In primary care, discuss concerns with your local consultant paediatrician hotline or the paediatric consultant on call the same day if there is a high index of suspicion regarding a possible abdominal tumour in a CYP.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	90.1% (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.

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**Discuss concerns over the telephone with the consultant paediatrician hotline or local equivalent service before referring a CYP from primary care in which differential diagnosis includes a possible tumour to ensure the CYP is seen within the most suitable timeframe (ideally within two weeks).**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	89.6% (Delphi Round 2)

### Rationale

A prolonged symptom interval with abdominal tumours 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<sup>[142]</sup>. 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<sup>[143, 144]</sup>. 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.

---

**Abdominal tumours can progress rapidly over days, increasing in size and causing pressure effects (e.g., breathing difficulties or bowel obstruction and ischaemia). If there is suspicion of an abdominal tumour this should be discussed in person 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	92.7% (Delphi Round 2)

### Rationale

The Guideline Development Group (GDG) and multi-disciplinary workshop members felt that there was low awareness of how abdominal tumours can progress rapidly and the urgency with which these children and young people need to be seen to ensure prompt diagnosis.

## Best practice – imaging

**Request an ultrasound of the abdomen and pelvis as the initial investigation for a CYP who has a suspected abdominal tumour. This should not delay referral from primary care.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	80.4% (Delphi Round 1)

### Rationale

The expert radiologists present at the multi-disciplinary workshop agreed that an ultrasound of the abdomen and pelvis was the initial investigation of choice for suspected abdominal tumours. “This should not delay referral from primary care” was added after the Delphi consensus based on the free text comments from some primary care clinicians who reported that they are not able to request paediatric investigations directly or that there are often long waits for imaging. It was felt that waiting for imaging should not delay referral from primary care. It is important to note here that a normal ultrasound with ongoing symptoms may warrant further imaging.

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**For abdominal tumours, imaging results should be interpreted by a professional with expertise and training in reporting ultrasound scans in children and young people.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

### 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 imaging in children should be interpreted by a healthcare professional with expertise in this area.

## Predisposing factors

Be aware that some predisposing factors such as a history of hemihypertrophy, WAGR (Wilm's tumor, aniridia, genito-urinary abnormalities, and mental retardation) syndrome, Denys Drash syndrome, hypospadias, cryptorchidism, aniridia, Beckwith Wiedemann or hereditary retinoblastoma, are associated with an increased risk of childhood abdominal tumours. 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	83.5% (Delphi Round 1)

### Rationale

The above are all associated with an increased risk of childhood abdominal tumours and therefore their presence should alert the clinician to this possibility and may lower their threshold for referral and investigation <sup>145-147</sup>.

## Presentation of a child with a potential abdominal tumour

Abdominal tumours encompass a large number of different tumours, namely Wilms' tumour, neuroblastoma, hepatoblastoma, abdominal lymphomas, soft tissue sarcomas and germ cell tumours.

The following symptoms and signs are all associated with childhood abdominal tumours. Their presence should alert the clinician to this possibility:

### Core symptoms

- Abdominal mass
- Abdominal pain
- Haematuria
- Abdominal distension

### Associated symptoms

- Weight loss
- Pallor/petechiae
- Fever
- Malaise
- Jaundice
- Hepatosplenomegaly
- Bone pain
- Neurological symptoms from spinal cord compression
- Bladder/bowel/erectile dysfunction due to spinal cord compression
- Urinary retention

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

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

The selection of core symptoms was based on data from meta-analyses and discussions at a multidisciplinary workshop. Weight loss, pallor/petechiae, fever, malaise, jaundice, hepatosplenomegaly, bone pain, neurological symptoms or bladder/bowel/erectile dysfunction from cord compression and urinary retention were also agreed by the Guideline Development Group (GDG) and the multi-disciplinary workshop members as associated symptoms.

The multidisciplinary workshop group agreed that whilst these symptoms were not individually as common, they were all signs that in combination with the main symptoms should warrant thought about malignancy in the abdomen.

## Assessment of a child with a potential abdominal tumour

### 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 an abdominal tumour.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	94.8% (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.

### Assessment

**Examine the abdomen, including external genitalia and hernia orifices, visualise and palpate the spine and perform a neurological examination in a CYP with signs/symptoms that may be due to an abdominal tumour.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	93.6% (Delphi Round 1)

### Rationale

Symptoms are usually progressive and can cause other seemingly unrelated symptoms and/or signs due to pressure on other structures, such as such as respiratory distress, spinal cord compression or bowel obstruction. It is important to fully assess a child presenting with symptoms suggestive of an abdominal tumour for any neurological deficits which if present would increase clinical suspicion and highlight the need for urgent referral to secondary care and imaging.

**Perform a urine dipstick to exclude infection and identify any protein or blood that would warrant referral to secondary care.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	89.7% (Delphi Round 1)

### Rationale

A common pitfall leading to delayed diagnosis can be incorrectly attributing symptoms to a urine infection because clinical presentations of abdominal tumours can be similar to presentations with urine infections. It is important to perform urinalysis on all children and young people presenting with abdominal pain to identify and treat an infection if present or identify any microscopic findings needing referral. All children should be followed up on completion of antibiotics, if symptoms persist, positive urinalysis findings persist, or diagnostic uncertainty exists, in which case children and young people should be referred to secondary care for further investigations.

## **Consider recording blood pressure if the correct size cuff is available to identify hypertension.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	74.2% (Delphi Round 1)

### **Rationale**

The multi-disciplinary team felt that whilst hypertension is a symptom of renal tumours in childhood, the blood pressure measurement is only useful if done using the correct cuff. General practitioners amongst the team felt that this is not often available within primary care and so this was phrased as a 'consider' option.

## **Be aware that weight loss can be a sign of an abdominal tumour. Measure weight and compare to any previous measurements in CYP with signs or symptoms suggestive of an abdominal tumour. Plot these measurements on age-appropriate growth charts if available to you to monitor change when reviewing symptoms.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	88.3% (Delphi Round 2)

### **Rationale**

The Guideline Development Group (GDG) felt that poor growth, especially weight loss is a worrying feature in addition to pain. As such, measuring and comparing the weight to previous is advised. Healthcare professionals should be aware the 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.

## Signs and symptoms of a child with a potential abdominal tumour

### Abdominal mass

**Abdominal masses due to a tumour can be in the abdominal cavity or retroperitoneal.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**Hepatosplenomegaly can also be present with an abdominal tumour.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, haematuria, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/ bladder/erectile dysfunction) in a CYP presenting with an abdominal mass.**

Strength of evidence	2++
Recommendation form	Conditional
Consensus achieved	99.0% (Delphi round 1)

**Be aware that diagnostic delay has been associated with failure to examine a child who presents with non-specific symptoms.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**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 when a CYP presents with refusal to weight bear.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	94.9% (Delphi Round 1)

### Rationale

The multi-disciplinary team could recall cases of delay in diagnosis where a child who was refusing to weight bear had not had an examination of their abdomen. As such it was felt important to highlight this.

## Diagnostic pitfalls

Be aware that delayed diagnosis has been associated with:

**Failure to perform an abdominal examination in a child who is distressed/crying. Offer to examine the child once they have settled or ask a colleague to perform the examination.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	94.8% (Delphi Round 1)

### Rationale

The guideline development 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:".

## Request ultrasound imaging for

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

**A CYP with a palpable abdominal mass (unless this is felt to be faeces, in which case a review of the CYP after disimpaction is important).**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	83.6% (Delphi round 1)

**A CYP with suspected hepatomegaly or splenomegaly on examination.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	83.5% (Delphi round 1)

## Abdominal Pain

**Abdominal pain is a common childhood presentation and in the majority of cases will not be due to an abdominal tumour.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**Ask about the presence of other symptoms of an abdominal tumour (haematuria, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with persistent abdominal pain (occurring on most days for a two-week period).**

Strength of evidence	2++
Recommendation form	Conditional
Consensus achieved	92.8% (Delphi round 1)

**Examine the abdomen in a CYP with abdominal pain to elicit any masses or hepatomegaly**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	99.0% (Delphi round 1)

**Be aware that if the tumour is retroperitoneal, the presenting complaint may be back pain. Examine the abdomen in a CYP presenting with back pain.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	95.9% (Delphi round 1)

## Rationale

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed that while abdominal pain is a very common childhood presentation, in most cases it is not due to a tumour. Each child with persistent pain should be assessed for the presence of any other symptoms or signs. Back pain is an unusual childhood presentation and a thorough abdominal examination is essential for any child presenting with back pain.

## Diagnostic pitfalls

Be aware that delayed diagnosis has been associated with:

**Attributing abdominal pain to constipation despite no improvement with laxatives. Assess response to laxatives by reviewing the CYP at regular intervals and taking a full history and examining their abdomen.**

Strength of evidence	3
Recommendation form	Conditional
Consensus achieved	87.7% (Delphi round 1)

### Rationale

The multidisciplinary team agreed that delayed diagnosis had been seen when abdominal pain had been presumed to be constipation despite no improvement following regular laxatives and/or successful disimpaction or lack of planned follow-up to review symptoms after treatment.

## Request ultrasound imaging for

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

**Persistent abdominal pain with one or more other symptoms that may be due to an abdominal tumour (abdominal distension, mass, haematuria, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction)**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	86.6% (Delphi round 1)

## Haematuria

### A renal tumour can present with macroscopic or microscopic haematuria

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

### Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, mass, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with haematuria in the absence of another known cause.

Strength of evidence	2++
Recommendation form	Strong
Consensus achieved	96.9% (Delphi round 1)

### Do a careful abdominal examination to elicit any potential retroperitoneal mass in all CYP with haematuria.

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	92.7% (Delphi round 1)

## Rationale

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed that persistent, unexplained macroscopic or microscopic haematuria warrants full assessment for the presence of any other symptoms or signs further investigation. The presence of additional features can help inform urgency of referral and investigations.

## Diagnostic pitfalls

Be aware that delayed diagnosis has been associated with:

**Attributing persistent haematuria to a UTI despite no response to antibiotics. Arrange to see the CYP again at the end of the course of antibiotics to assess the response. If there is no response, consider discussion with paediatrician for advice.**

Strength of evidence	3
Recommendation form	Conditional
Consensus achieved	82.4% (Delphi round 1)

## Request ultrasound imaging for

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

### Unexplained persistent (occurring for two weeks or more) macroscopic haematuria.

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	87.7% (Delphi round 1)

### Haematuria plus one or more other symptoms that may be due to an abdominal tumour (abdominal pain, abdominal distension, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction).

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	87.6% (Delphi round 1)

## Abdominal Distension

**Abdominal distension is incredibly common in childhood, especially in pre-school children. In most cases abdominal distension is not due to an abdominal tumour.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

### Rationale

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, haematuria, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with an abdominal distension.**

Strength of evidence	2++
Recommendation form	Strong
Consensus achieved	97.9% (Delphi round 1)

**Examine the abdomen, spine and peripheral nervous system examination in a child or young person presenting with abdominal distension.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	Not applicable

### Rationale

The multidisciplinary workshop group agreed these statements. The GDG felt they should be highlighted but did not need to go out for consensus.

**Be aware that abdominal distension caused by a tumour will not fluctuate but will increase in size progressively over time. Arrange another appointment to review the symptoms and re-examine the CYP.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	83.5% (Delphi round 1)

### Rationale

The multidisciplinary workshop group and the Guideline Development Group (GDG) agreed that abdominal distension should be followed up closely in children to distinguish those due to tumour from more common benign conditions. The identification of progressive distension, development of additional symptoms at any of these reviews or the failure to improve despite seemingly appropriate treatment should be referred to secondary care.

## Diagnostic pitfalls

**Be aware that delayed diagnosis has been associated with:**

**Attributing abdominal distension to constipation despite no improvement with laxative treatment. Assess response to treatment in these CYP by seeing them at regular intervals.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	84.5% (Delphi round 1)

**Failure to examine the abdomen at review of a CYP on treatment for constipation. Palpate the abdomen when reviewing CYP to assess response to treatment.**

Strength of evidence	4
Recommendation form	Conditional
Consensus achieved	94.9% (Delphi round 1)

## Request ultrasound imaging for

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

**A CYP with rapidly increasing abdominal distension. This should not delay referral from primary care.**

Strength of evidence	4
Recommendation form	Strong
Consensus achieved	83.5% (Delphi round 1)

# Implementation Strategy and Future Work

## Guideline implementation

The guideline implementation will be supported with the launch of the Child Cancer Smart awareness campaign ([www.cclg.org.uk/childcancersmart](http://www.cclg.org.uk/childcancersmart)) including clinical guidelines, quick reference and educational package 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.

## Further 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 or changes in referral pathways, then a full revision will be conducted in line with AGREE II criteria.

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## Summary of Recommendations

### Diagnosis of abdominal tumours in children and young people

A summary for healthcare professionals

#### Consider an abdominal tumour in any child presenting with:

- Abdominal mass
- Abdominal pain
- Haematuria
- Abdominal distension

#### Associated symptoms:

- Weight loss
- Pallor/petechiae
- Fever
- Malaise
- Jaundice
- Hepatosplenomegaly
- Bone pain
- Neurological symptoms

#### Ask about common predisposing factors

- Hemihypertrophy
- WAGR syndrome
- Denys Drash syndrome
- Hypospadias
- Cryptorchidism
- Aniridia
- Beckwith Wiedemann
- Hereditary retinoblastoma

#### Assess with:

##### History:

- Associated symptoms
- Any predisposing factors
- Family history

##### Examination of:

- Abdomen
- Spine
- Weight
- Neurological examination
- Urine dipstick
- Blood pressure

#### 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

- High risk of tumour – SAME DAY referral to secondary care
- Lower risk\* - discuss with paediatrics via telephone to advise best route to be seen

#### Imaging

- High risk of tumour: URGENT ultrasound imaging
- Lower risk\*: ultrasound imaging within 4 weeks

\*abdominal tumour in differential diagnosis, low Index of suspicion

#### Abdominal mass

- Abdominal masses due to a tumour may be in the retroperitoneum.
- Hepatosplenomegaly may also be present with an abdominal tumour.
- Ask about the presence of other symptoms (abdominal pain, haematuria, abdominal distension, malaise, jaundice, bone pain, neurological symptoms, erectile dysfunction).
- Be aware that diagnostic delay has been reported in a child who presents with non-specific symptoms.
- Be aware that abdominal masses can cause pressure on the spinal cord. Such pressure may present as “off legs” or to refuse to weight bear. Examine the young person presents with refusal to walk.

#### Ultrasound imaging required:

- A palpable abdominal mass
- Suspected hepatomegaly or splenomegaly

#### Diagnostic pitfalls:

- Failure to perform a thorough abdominal examination (distended/crying)
- Failure to examine the abdomen when a child is weight bear or walk

#### Abdominal pain

- Abdominal pain is a common childhood symptom. Most cases will not be due to an abdominal tumour.
- Ask about the presence of the other symptoms (haematuria, abdominal distension, weight loss, bone pain, neurological symptoms and hepatomegaly).
- Examine the abdomen in a CYP with abdominal pain or hepatomegaly.
- Be aware that if the tumour is retroperitoneal, it may not be back pain. Examine the abdomen in a CYP.

#### Ultrasound imaging required:

- Persistent abdominal pain WITH one or more of the above symptoms or an abdominal tumour

#### Diagnostic pitfalls:

- Attributing abdominal pain to constipation and giving laxatives. Assess response to laxatives before repeating intervals and taking a full history and examination.
- Failing to examine the abdomen in a child with abdominal back pain

\* Persistent = present on most days for more than 2 weeks



may arise either within the abdominal  
cavity or in association with an  
injury. Symptoms of an abdominal tumour  
include abdominal distension, weight loss, fever,  
neurological symptoms, and bowel, bladder or  
urinary symptoms. A child may be  
linked to a failure to examine a  
child's abdomen. Use neurological symptoms due to  
a tumour may lead children to present as  
neurological symptoms. Examine the abdomen when a child or  
young person presents with back pain.

usually  
abdominal examination in a baby who is  
ill. If a child has presented with refusal to

presentation and in the majority of  
cases of an abdominal tumour  
symptoms of an abdominal tumour  
include weight loss, fever, malaise, jaundice,  
neurological symptoms and bowel/bladder/  
erectile dysfunction) abdominal pain to elicit any masses  
retroperitoneal, the presenting complaint may  
be back pain. Examine the abdomen when a CYP presenting with back pain.

more other symptoms attributable to  
a tumour despite no improvement with  
antibiotics. Review the CYP at regular  
intervals by reviewing the CYP at regular  
intervals examining their abdomen.  
Examine the abdomen when a child or  
young person presents with back pain.

weeks

## Haematuria

- A renal tumour can present with macroscopic or microscopic haematuria
- Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, mass, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction) in a CYP presenting with haematuria in the absence of another known cause.
- Do a careful abdominal examination to elicit any potential retroperitoneal mass.
- Follow local haematuria in children guideline to support clinical decision making

### Ultrasound imaging required:

- Unexplained persistent macroscopic haematuria
- Haematuria WITH one or more other symptoms attributable to an abdominal tumour

### Diagnostic pitfalls:

- Attributing persistent haematuria to a UTI despite no improvement with antibiotics

\* Persistent = present on most days for more than 2 weeks

## Abdominal distension

- Abdominal distension is incredibly common in childhood, especially in pre-school children and in most cases is not due to an abdominal tumour
- Ask about the presence of the other symptoms of an abdominal tumour (abdominal pain, haematuria, weight loss, fever, malaise, jaundice, bone pain, neurological symptoms and bowel/bladder/erectile dysfunction)
- Examine the abdomen, spine and peripheral nervous system examination in a child or young person presenting with abdominal distension.
- Abdominal tumours causing abdominal distension tend to progress and does not improve.

### Ultrasound imaging required:

- Increasing abdominal distension
- Abdominal distension with one or more other symptoms attributable to an abdominal tumour.

### Diagnostic pitfalls:

- Attributing distension to constipation despite no improvement with laxatives
- Failure to examine the abdomen at review of a child with constipation

\*Persistent = present for more than 2 weeks

[www.cclg.org.uk/childcancersmart/abdominal](http://www.cclg.org.uk/childcancersmart/abdominal)

## GDG members, multidisciplinary workshop and Delphi participants

### GDG Members

1. Dr Shaarna Shanmugavadivel (CCLG Early Diagnosis Fellow and Child Cancer Smart Lead, University of Nottingham, Nottingham, UK)
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4. Dr Sophie Wilne (Consultant Paediatric Oncologist, Nottingham University Hospitals NHS Trust, Nottingham, UK)
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16. Dr Kate Bishton
17. Dr Sian Kenny
18. Dr Penny Edrich
19. Dr Julia Foley
20. Dr Laura Hannaford
21. Dr Hannah Green
22. Dr Emma Goldstein
23. Dr Joanne Sinclair
24. Dr Samantha Robinson
25. Dr Claire Grimstvedt
26. Dr Teresa Tang
27. Dr Lucy Follis
28. Dr Katherine Teare
29. Dr Laura Maher
30. Dr Sonja Adhiyaman
31. Dr Anum Alamgir
32. Dr Zoe Hunter
33. Dr Bethany Thomas
34. Dr Sarah Cousins
35. Dr Amber Nielsen
36. Dr Verity Lowndes-Burt
37. Dr Christine Clayton
38. Dr Sarah Arthur
39. Dr Kay Jones
40. Dr Hannah Williams
41. Dr Sana Shahid
42. Dr Alison Lee
43. Dr Janet McIlpatrick
44. Dr Rachael King
45. Dr Chiranthi Siriwardena
46. Dr Alexandra Peake
47. Dr Harriet Williamson
48. Dr Pamela Ross
49. Dr Marie Cohen
50. Dr Toni Hazell
51. Dr Elizabeth Lacey
52. Dr Laura Edwards
53. Dr Ella Bosmith
54. Dr Elizabeth Field
55. Dr Christine Gray
56. Dr Clare Nwosu
57. Dr Esmat Abbas

## 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.

### Grades 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)<sup>3</sup>

Judgement	Recommendation
Undesirable consequences clearly outweigh desirable consequences	Strong recommendation against.
Undesirable consequences probably outweigh desirable consequences	Conditional recommendation against.
Balance between desirable and undesirable consequences is closely balanced or uncertain	Recommendation for research and possible conditional recommendation for use restricted to trials.
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 eg on the quality or size of the evidence base, or patient preferences. Conditional recommendations should include ‘should be considered’ in the wording.



## Are you Child Cancer Smart?

Child Cancer Smart is an evidence-based public and professional awareness campaign to improve early diagnosis of cancer in children and teenagers aged 0-18.

The campaign recommends that if a child has had:



**3**

persistent  
symptoms

or



**3**

visits to  
the doctor

or



**3**

weeks of  
unexplained  
symptoms

their doctor should pick up the phone and call the local on-call paediatrician.

# Early diagnosis can save lives.

Find out more at [www.cclg.org.uk/childcancersmart](http://www.cclg.org.uk/childcancersmart)

## Why 3 symptoms, 3 visits or 3 weeks?

This message for professionals has been chosen based on the evidence we have. We know that some symptoms of cancer are more obvious than others and so whilst, for some, cancer will be considered very quickly, for others it may take longer.

In the Childhood Cancer Diagnosis (CCD) study, 50% of solid tumours were diagnosed within 3 weeks of seeing a healthcare professional, and leukaemias often much more quickly than that.

We want to shorten the time for the other 50%. Ensuring healthcare professionals are considering cancer in children at the 3-week mark if symptoms are progressive and unexplained will help us reach our target of diagnosing 75% of children and teenagers within 3 weeks from first contact with a healthcare professional.

## Let us know what you think, or get involved

If you've got any feedback on the Child Cancer Smart campaign, we'd love to hear it.

If you would like to get involved and support the Child Cancer Smart campaign - whether by contributing to the guideline development process, acting as one of our clinical champions, or in many other ways, we would be delighted to hear from you.

Contact us by completing the form at [www.cclg.org.uk/childcancersmart/your-thoughts](http://www.cclg.org.uk/childcancersmart/your-thoughts)





## The Children & Young People's Cancer Association

FUNDED BY

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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.