Update on the UMBRELLA study & associated Projects

- From 2019, the UMBRELLA study has been implemented as an amendment to the IMPORT study across the UK. The national sponsor continues to be GOSH and the international coordination is under the auspices of the SIOP Renal Tumours Study Group (SIOP-RTSG).
- Despite temporary halts to recruitment due to COVID19, the international study has registered over 581 patients from 17 countries, with some of these countries joining the study for the first time. The UK has recruited 80 patients. The first report has been shared with the RTSG Steering Committee.
- On the 27th Feb 2020 we launched our newly funded project ‘The Little Princess Wilms Tumour Knowledge Bank’. It is a collaboration between UCL/GOSH and the Sanger Centre in Cambridge, PI: Dr Sam Behjati.
- To date, Whole Genome Sequencing (WGS) has been performed on 250 Wilms tumours collected in the UK and Germany with a focus on metastatic cases and infants diagnosed under the age of 12 months. A further 80 WT will have WGS, with results expected in Spring 2021.
- Despite the delays caused by lockdown, we also managed to continue two projects to assess circulating molecular biomarkers:
  1. a liquid biopsy study (whole exome sequencing on circulating tumour DNA from plasma of WT patients) in conjunction with Dr Gudrun Schleiermacher, Paris
  2. Analysis of microRNAs in diagnostic serum, with Dr Matt Murray, Cambridge.

The Little Princess Wilms Tumour Knowledge Bank

On the 27th Feb 2020, we launched our newly funded project ‘The Little Princess Wilms Tumour Knowledge Bank’ which has been named after the charity that kindly donated the funds. Dr Sam Behjati hosted the project launch meeting at the Sanger Centre, Cambridge where the LPT charity founders, CEO, the study Chief Investigator Prof Kathy Pritchard-Jones and a group of clinicians and scientists from UCL Great Ormond Street Institute of Child Health attended and discussed the plans to start this collaboration.

Updated CCLG clinical guidelines

The clinical guidelines that accompany the amendment of IMPORT into the UMBRELLA study explain the changes that have updated our previous standard of care. These are based on the SIOP WT 2001 randomised trial and aim to incorporate new evidence from international partners on significance of tumour volume after pre-op chemo, as well as definitions of metastatic disease. There is also an agreed international standard approach to treatment of relapsed Wilms tumour that incorporates our national experience from UKWR. The updated guidelines are published on the CCLG website ‘members’ area’.

UK PPIE meeting, 6th Feb 2020

Prof Kathy Pritchard-Jones, Mr Max Pachl (paediatric surgeon) and Mrs Reem Al-Saadi (study manager) met with the UK patient and parent group, led by Mrs Angela Polanco (PPIE lead) in Birmingham to plan new projects. We discussed the importance of collecting long term outcome data and have taken into account their views and priorities. We talked about linking clinical trial data to national cancer registries and other routine health care data to aid long term outcomes research. This was seen to both add value to the original trials as well as being an important research priority in itself.
Recent Abstracts

- **SIOP 2020 presented abstracts:**
  - Variation in adherence to protocol recommendations for pre- and post-operative treatment of children with Wilms tumour in two consecutive studies in the UK and Ireland (oral, S Tugnait).
  - Wilms tumours with intravascular extension in the UK and Ireland IMPORT study: reasons for stage III (poster, K Dzhuma).
- An update on the UMBRELLA study was presented during the CCLG Winter meeting on the 27-28 January 2020, and a trials update was presented during the NCRI trials meeting on the 29th January 2020
- Poster at CCLG conference Jan 2020: S Tugnait et al, Review of chemotherapy delivery for Wilms tumours in the UK and Ireland in the SiOP WT 2001 and IMPORT studies

**RTSG General Assembly & Brazilian PPIE Meeting**

The annual SIOP RTSG GA took place in Rio de Janeiro on 11-13th March 2020, hosted by Dr Beatriz De Camargo & Dr Joaquim Caetano Aguirre Neto for the Brazilian WT group. Five UK delegates attended the meeting which also included a Brazilian parents and survivors group meeting.

During the PPIE meeting we linked with Mrs Angela Polanco (Founder of Bethany’s Wish) via teleconference for her input of her experience and the UK perspective, which was warmly welcomed. It was great to meet the group in person; which included 2 survivors in their 30s now, one of who attended with her baby. The Brazilian group met afterwards virtually and is planning to hold monthly follow meetings virtually in 2021. They aim to attract more parents, develop new initiatives and support research.

**Survivor and now Researcher donates hair to LPT**

Suzi Tugnait, a medical student and research assistant in the Wilms team, was five-years-old when she underwent chemotherapy which led her to lose her hair. At the time she struggled to find a wig, therefore Suzi said she was delighted to be able to help The Little Princess Trust by donating her long locks. The charity will use these to make a wig for a young person who has lost their own hair. Friends and family also sponsored her big cut – which has so far raised more than £3,000 for LPT.
Thank you to all our supporters, and especially to the charities Bethany’s Wish and The Little Princess Trust.

Stay safe!
Recent Publications

CCLG Parent’s Guide Factsheet: Wilms Tumour (updated 03/06/2020)

Scientific Papers published in 2020


LAY SUMMARIES OF PUBLICATIONS

1. WILMS TUMORS AND THEIR PRECURSORS: RADIOLOGICAL DIAGNOSIS VERSUS HISTOLOGY


**Background:** This research addresses a difficult issue in clinical practice regarding the diagnosis of patients with multiple renal nodules. The aim of this study was to compare the imaging features of renal nodules with their final histology and identify radiological factors that would better differentiate Wilms tumors (WT) from nephrogenic rests (NR) - benign lesions from which Wilms tumors may arise.

**Methods:** Images of the patients with bilateral and unilateral multiple renal tumors were reviewed by a senior radiologist blinded to histology findings. All nodules were classified as Wilms tumors or nephrogenic rests according to the radiological criteria.

**Results:** 175 nodules were resected after a median of five courses of chemotherapy for the first side and 13 courses for the second side. The radiology/pathology correlation showed 118 (74.6%) nodules adequately classified, 20 (11.3%) false positive for WT (9 patients) and 20 (11.3%) false negative (9 patients). WTs were significantly bigger at diagnosis and preoperative assessment and their size decreased quicker than NR (p<0.0001) under chemotherapy. WT and NR had a similar homogeneity rate. Only 5% of nodules thought NR that were finally WT were heterogeneous while 17% of nodules initially diagnosed as WT that were finally NR were homogeneous.

**Conclusion:** Radiological misdiagnosis occurred in 25% of kidney nodules. The size of nodules at diagnosis and their decrease under neoadjuvant chemotherapy may be a helpful criterion recognizing WT from NR before surgery. Other criteria like homogeneity did not seem key features and lost value in small lesions.

For more lay summaries please email us at:  
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