ESPGHAN
European Society for Paediatric
Gastroenterology, Hepatology and Nutrition

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Rue De-Candolle 16
1205 Genève, Switzerland
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Working Group - Report for Gastroenterology Committee Strategy Day

Working Group Title: Paediatric Polyposis Working Group

(5th Sept 21; Author Warren Hyer)

Working Group Members and Mandates

Name	Date of entry	1st Term	Positions held within WG (e.g. Secretary treasurer etc.)	Dates held of these positions
Warren Hyer	2015	2015	Co chair and convenor	2015 - current
Shlomi Cohen	2015	2015	Co- chair	2015- current
Andrew Latchford	2016	2016	InSight representative and guideline advisor	2016- current
Carol Durno	2017	2017	NASPGHAN representative and guideline advisor	2017
Thomas Attard	2017	2017	NASPGHAN representative and core group	2017
Marcus Auth	2016	2016	Core group member	2016
Mike Thomson	2015	2015	Endoscopy Working group lead and WG advisor	2015
Jerome Viala	2015	2015	Member, contributor	2015
Cecile Talbotec	2015	2015	Member, contributor	2015

Victor Vila Miravet	2015	2015	Member, contributor	2015
Corina Pienar	2015	2015	Member, contributor	2015
Seth Septer	2015	2015	Member, contributor and lead for paediatric endoscopy	2015
Emmanuel Mas	2015	2015	Member, contributor and elected representative on gastro GIC	2015
Johannes Spalinger	2015	2015	Member, contributor	2015
Jackie hawkins	2016	2016	Nurse contributor. Patient advocate	2016

Summary of Activities in 2020/21

A. WG meetings (date and any main points/activities)

Meeting	Planned Working group meeting, ESPGHAN WCPGHAN 2021; all members. Face to face cancelled – moved instead to a CORE group meeting regarding planned research projects
Location	WCPGHAN
Date	May 2021 – cancelled, no WG meetings at WCPGHAN
Main Points / Activities	Planned meeting to update the WG regarding planned research projects allocated to teams. Role and management of a future polyposis registry specifically seeking data for rare but significant implications of paediatric polyposis

Meeting	Core group authors, WH,SC, AL, TA, CD
Location	On line video conference
Date	Jan 2021
Main Points / Activities	• Identify, discuss and then formalize the direction of research for 2-5 year. • Incorporating NASPGHAN experts to create an international selection of experts, each with large cohorts of polyposis patients, with previous publication expertise in the area of paediatric polyposis. • 5 Projects identified, dissemination of projects to the WG. • Grant secured ESPGHAN for database (REDCAP Israel 2021)
Outcome of the core	Developing dataset for the clinical queries. Close to conclusion and ready to go live Autumn 2021 1) To evaluate the long term prognosis of children with JPS in the absence of a known germline mutation (*SMAD4, BMPR1A, PTEN, ENG*) *Approx prevalence 1:150,000* 2) To evaluate the optimal size of a PJS polyp that predicts intussusception; looking at: -children who have small bowel polyps and no intervention, -children who present with intussusception - children who have elective polypectomy to prevent intussusception *Approx prevalence – 1:200,000* 3) To evaluate the outcomes for patients with combined *BMPR1A*, with *PTEN* mutation. *Prevalence unknown* 4) To evaluate paediatric manifestations of desmoid disease in paediatric FAP *Prevalence unknown*

Meeting	Video conference with NAPSGHAN leads in polyposis
Location	Video on line
Date	May 2021
Main Points / Activities	Enrollment and support by NASPGHAN team for the REDCAP database to approach the clinical issues explored by the ESPGHAN WG. Broad and widespread support for this project.

B. Educational activities

(i) Educational Events

Presentation planned at the ESPGHAN summer school (cancelled with COVID).
Currently no plans for face to face educational events until COVID crisis has passed.

There is an opportunity for an on line one day workshop on Management of Paediatric Polyposis for early 2022. The current increased use of video conferencing and improvement in acceptability of on line live video learning lends itself to this educational programme. The WG are rapidly considering this as an ideal training and learning opportunity for the WG, and ESPGHAN/NASPGHAN.

C. WG Publications / Guidelines / Position Papers

(i) Completed/near completion

Publication ID *(to be filled in by Office)*	
Full Publication Title	ACG Clinical Report and Recommendations on Transition of Care (TOC) in Children and Adolescents with Hereditary Polyposis Syndromes
Authors	Attard et al
Publication Type	☐ Position Paper
Journal, Year, Volume, Page Numbers	S Published 2020
More information if available	This guidance has been led by Tom Attard on behalf of the WG through the American College planning transition of care from childhood to adult care providing clear guidance of clinicians. The role of transition of care had been highlighted in the position papers published March 2019 – this is the first paper to be published specifically looking at Best Practice for TOC

(ii) future topics considered in 2020/21

International Registry of Pediatric Polyposis syndromes to assess the clinical questions raised below. An online database and analysis platform to study pediatric polyposis syndromes - International multi-center research

Proposers:

- Prof Shlomi Cohen, Dana Dwek Childrens Hospital, Tel Aviv;
- Dr Warren Hyer; Paediatric Lead, St Mark's Hospital Polyposis Registry;
- On behalf of the Paediatric Polyposis Working Group ESPGHAN.
- Ready to go live.
- Meeting with SC and WH face to face again cancelled on 3 sequential occasions with travel restrictions; COVID 19.

Primary objectives	Primary endpoints
To evaluate the long term prognosis of children with JPS in the absence of a known germline mutation (*SMAD4, BMPR1A, PTEN, ENG*) *Approx prevalence 1:150,000*	• Comparison of the number of polyps, symptoms and surveillance between the 2 groups (with and without known germline mutation.)
To evaluate the optimal size of a PJS polyp that predicts intussusception; looking at: 1)children who have small bowel polyps and no intervention, 2)children who present with intussusception 3) children who have elective polypectomy to prevent intussusception *Approx prevalence – 1:200,000*	• Comparison of compare the size of the polyp in the 3 groups and compare against patient size leading to evidence about the size of polyp requiring polypectomy to avoid intussusception
To evaluate the outcomes for patients with combined *BMPR1A*, with *PTEN* mutation. *Prevalence unknown*	• Comparison of phenotype of those patients with combined *BMPR1A /PTEN* mutation to those with only *PTEN* or *BMPR1A* mutation.
To evaluate paediatric manifestations of desmoid disease in paediatric FAP *Prevalence unknown*	• Description of this subgroup of patients.

D. Financial Report / Budget for 2020: Will be added by EPGHAN Office

Application for ESPGHAN grant submitted July 2021

PROPOSED BUDGET INCLUDING A DETAILED DESCRIPTION OF THE GRANT COORDINATION AND DISTRIBUTION AMONGST CENTRES

No budget for 2020. No expenses incurred

Additional comments on financial report:

Application for ESPGHAN Grant made for Registry July 2020 – accepted project in progress

Category	**Amount (€)**
Technical costs of informatics: building the retrospective questionnaire and maintenance of the database	10,000
Study technical coordinator (part-time)	5,000
Coordinating Centre	5,000
Participating centres (ethics committee fees, admin)	10,000
Overhead (Tel-Aviv Souraski Medical Center) 15%	4,500
Meetings	9,000
Sum :	43,500

Outlook on Planned Activity for 2020/21

A. Working Group meetings (date and any main points/activities)

Meeting	Core Group meeting; WH and SC
Location	Dana Dwek Childrens Hospital
Date	Scheduled face to face October 2021; again cancelled. Move to video meet
Main Points / Activities	Face to face meeting meeting between Cohen and Hyer to finalise the protocols for clinical studies listed below. Plan an online one day workshop for Europe and beyond – with the support and promotion of ESPGHAN

Proposed Educational activities

(i) E-Learning

Full Project Name	One day workshop for trainees and experienced paediatric gastroenterologist on Polyposis Syndromes in children; and specifically on safe polypectomy in children and adolescents
Project Timeline	Finalise Oct 2021, with proposal to ESPGHAN late 2021
Project Leader	Cohen and Hyer and Attard
more information if available (applied for UEG support? Etc)	One day event. Video conference under the auspices of ESPGHAN. Clear educational objectives. Endoscopy video learning on safe polypectomy

B. Proposed Publications

Work in progress:

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C. Financial Report / Budget for 2020:

- **Kindly attach or copy paste your submitted budget request for 2021**

No current costs incurred by non ESPGHAN members invited to attend or present at ESPGHAN 2021 (see below)

No proposed costs for 2022 other than application for ESPGHAN funds for a ESPGHAN polyposis registry – grant application separate to this report. .