

**2012 SPP Spring Meeting
Vancouver, BC, Canada
September 29-October 2, 2011**

Meeting Needs Assessment

The practice of pediatric pathology requires up to date knowledge of the diseases affecting children, including their scientific basis, clinical spectrum, pathologic classification, and current research activities. The Society for Pediatric Pathology Annual Meeting is intended as an ongoing resource to meet the educational needs of pediatric pathologists, general pathologists whose practice includes pediatric pathology, pediatric pathology fellows, and pathology residents.

Meeting Objectives

Upon completion of this meeting, participants should be able to:

- Through the platform and poster scientific sessions, discuss current clinical problems and research activities in pediatric pathology.
- Through the symposium, discuss the basic science, pathologic and clinical aspects of a specific topic.
- Through the workshops, apply practical instruction from recognized expert pathologists in specific areas of pediatric pathology to improve one's diagnostic skills and practice.

Synopsis

The 2012 SPP Spring Symposium will provide a comprehensive update on Hirschsprung disease. Specific topics to be included are the Embryology and Genetics, Preoperative Diagnosis by Rectal Biopsy, and Role of the Pathologist in the Intra- and Post-operative Management. Four specialists in the field have agreed to lecture at the Symposium: Drs. Robert O. Heuckeroth, Miguel Reyes-Mugica, Jacob Langer, and Raj Kapur.

Symposium Needs Assessment

Hirschsprung disease is a congenital malformation that affects 1:5000 infants and is an important diagnostic consideration in surgical and forensic pathology. Accurate diagnosis of the disorder has important implications, because it commits the patient to a surgical procedure, confers an increased likelihood of co-incident malformations in other organ systems, and confers an increased risk of Hirschsprung disease or related anomalies in family members. Much new information has been learned about the genetics, embryology and pathology of this condition over the last decade, but the information is complex, and its practical implications are unfamiliar to most pathologists. As a consequence, significant knowledge gaps exist, particularly in regard to practice guidelines for procurement, handling, assessment, and reporting of suction rectal biopsies; use of ancillary methods to facilitate diagnosis; the role of intraoperative frozen sections in surgical management, application and interpretation of molecular genetic tests; and the pathological findings that correlate/predict post-operative complications. Existence of these knowledge gaps is evident from frequent postings to the pediatric pathology listserv, (a free standing, University of Washington pediatric pathology based and maintained, highly respected and avidly used resource for academic and private practice pediatric pathologists and their colleagues). The perceived need for more guidance in this area was formally recognized by the SPP Practice Committee and discussed by the SPP Council, in 2007 (see SPP Newsletter dated August, 2007; volume 43 #1). An informal poll of the SPP Education Committee members indicated a strong, needs based assessment for clearer distinction of the complex pathologies of HD and its associations, and intestinal neuronal dysplasia. The poll and the ongoing queries to the pediatric pathology listserv indicated address of HD and its differential diagnosis are critical to the pathologist's role in diagnosis and improved direction of patient care and patient outcomes for the reasons discussed below.

Research from the last decade has established that most, if not all, instances of Hirschsprung disease have a genetic basis, but multiple genes act as risk factors and combinatorial genetic and environmental effects exist. Although genotype-phenotype correlations exist, the penetrance of intestinal aganglionosis is variable and difficult to predict based on mutational analysis. Some specific genetic defects observed in Hirschsprung disease patients are associated with other malformations, the occurrences of which are explainable based on the nature of these genes' products and their roles during embryogenesis. In addition, some genetic alterations are associated with neoplasia or other disease processes, which may appear later in life. Pathologists' knowledge of these associations, their genetic bases, and the benefits/limitations of contemporary mutational analyses are critical, since pathologists direct and interpret relevant laboratory tests.

Diagnosis of Hirschsprung disease from suction rectal biopsies is the norm. However, approaches to the acquisition, handling and interpretation of these biopsies vary considerably, despite published protocols for sampling and histopathological evaluation of suction rectal biopsies. Pathologists need to be made aware of the many variables that influence the adequacy and interpretation of suction rectal biopsies, and approaches that can be used to improve the quality and accuracy of diagnoses. Use of special stains (e.g., acetyl-cholinesterase (AChE) histochemistry) in the work-up of rectal biopsies is particularly controversial, as some institutions regard them as essential, others use them as ancillary, and still others do not apply them at all. In the past three years, calretinin immunohistochemistry has been introduced as a potential ancillary method to facilitate diagnosis, and has replaced AChE in some laboratories. However, many pathologists are unaware of the benefits and limitations of calretinin staining, and/or how it might be integrated into their practice. Another significant challenge concerns diagnosis of conditions that might clinically mimic Hirschsprung disease (e.g., hypoganglionosis, visceral myopathy) and the role, if any, of suction rectal biopsies in their diagnosis. Among the latter, intestinal neuronal dysplasia may be the most controversial, since it is a histopathological phenotype embraced by many physicians (especially surgeons) as a surgically treatable, primary cause of intestinal dysmotility but discounted by others, as either a normal variant, or secondary phenomenon without established pathophysiological significance. Exclusion of intestinal neuronal dysplasia is a frequent reason for outside consultation (RK, personal experience), largely because the diagnostic criteria and their significance are not understood by most pathologists or clinicians.

The treatment for Hirschsprung disease is surgical resection of the aganglionic bowel and eventual anastomosis of proximal "euganglionic" bowel to the anus. Several different types of surgical anastomoses have been devised, each with its own theoretical advantages and disadvantages. Regardless of the approach, rates of post-operative complications, including persistent obstructive symptomatology are significant, and some patients require additional diagnostic biopsies and/or surgical procedures to manage their dysmotility. Several possible explanations for persistent post-operative symptoms have been suggested. Among these is "transitional zone pull-through", which refers to inadequate resection of neuroanatomically abnormal bowel that resides between the ganglion cell containing and aganglionic segments. Unfortunately, anatomic criteria, used to identify the transitional zone and methods to ensure its complete resection, are not well established. Moreover, conflicting information exists in the pediatric surgery literature regarding the effectiveness of longer vs. shorter resections of ganglionic bowel and their impact on post-operative outcomes. Pathologists need to be educated as to what is known about the transitional zone, how it might be assessed intraoperatively and post-operatively, and how to work with the surgeon to evaluate patients with post-operative obstructive symptoms. After a critical review of literature related to the transitional zone, it is possible to provide guidelines for the practicing pathologist, and improve upon the standard of specimen analysis currently practiced at many institutions, and, thereby, likely improve the care and outcomes for many patients.

Our slate of internationally recognized speakers will address all of these topics cognizant of the needs of the practicing pathologist. An effort will be made to present only evidence-based recommendations and highlight gaps that remain to be addressed in the future. The talks will be coordinated to complement one another and avoid significant overlap.

Symposium Objectives

Upon completion of this symposium, participants should be able to:

- Discuss the molecular genetic and cellular factors that influence enteric neurodevelopment, how disturbances in these factors contribute to the pathogenesis of congenital disorders of the enteric nervous system, and apply this knowledge to the diagnosis and management of patients with Hirschsprung disease.
- Apply an algorithm for the handling and interpretation (including ancillary histochemical and immunohistochemical methods) of suction rectal biopsies to exclude/diagnose Hirschsprung disease and related disorders.
- Recognize common surgical approaches to Hirschsprung disease and the intra-operative and conduct appropriate post-operative analyses to optimize patient management.
- Describe the histopathological correlates of the transitional zone in congenital aganglionosis, potential problems that result from a transitional zone pull-through, and how to avoid and diagnose/treat such complications.

Planner and Faculty Disclosure

As a sponsor accredited by the Accreditation Council for Continuing Medical Education (ACCME), the Society for Pediatric Pathology (SPP) must ensure balance, independence, objectivity, and scientific rigor in all its individually or jointly sponsored educational activities. All planners and faculty (speakers, authors and presenters) who participate in the planning and execution of an SPP educational activity are required to disclose whether they or their spouse/partner have any significant financial interest or other relationship (1) with the manufacturer(s) of any commercial product(s) and/or provider(s) of commercial services discussed in an educational presentation and (2) with any commercial supporters of the activity. (A “significant financial interest or other relationship” can include such things as grants or research support, status as an employee, consultant, major stock holder, member of speakers bureau, etc.) This applies to current relationships as well as any within the past twelve months. The intent of this disclosure is not to prevent a faculty member with a significant financial or other relationship from making a presentation. Disclosure is required so that planners may reasonably decide whether to make adjustments in the program and its faculty, and so that participants in the activity may formulate their own judgments as to whether the faculty member’s interests or relationships influenced the presentation with regard to exposition or conclusion. The SPP has also required that faculty disclose any products that are not labeled for the use under discussion and that the disclosure be made to the audience at the time of presentation.

All planners and faculty who participate in the planning and execution of an SPP educational activity have been informed of the SPP Policy on Disclosure of Relevant Conflicts of Interest. Faculty who have disclosed industry relationships have been counseled, in writing, of the necessity to deliver their presentations in a manner free of commercial bias. Written notification is made, in accordance with SPP Policy, to resolve any relevant conflicts of interest.

CERTIFICATE OF CME/SAM CREDIT OR PARTICIPATION

Accreditation Statement

The Society for Pediatric Pathology is accredited by the Accreditation Council for Continuing Medical Education (ACCME) to provide continuing medical education for physicians.

AMA Credit Designation Statement

The Society for Pediatric Pathology designates this live activity for a maximum of 15.25 *AMA PRA Category 1 Credit(s)*[™]. Physicians should only claim the credit commensurate with the extent of their participation in the activity.

International Physicians

The American Medical Association has determined that physicians not licensed in the US who participate in this CME activity are eligible for *AMA PRA Category 1 Credit(s)*[™].

Health Professionals

Health Professional participants (including residents and fellows-in-training) may claim hours to receive a Certificate of Participation for an activity designated for *AMA PRA Category 1 Credit(s)*[™].

CME Credits

Certificates of continuing medical education *AMA PRA Category 1 Credit(s)*[™] will be issued through the Society for Pediatric Pathology. CME credits will only be awarded after completion of an online evaluation form.

Self-Assessment Module Credits

The SPP is accredited by the American Board of Pathology to offer Self-Assessment Module (SAM) credits for the purpose of meeting the American Board of Pathology requirements for Maintenance of Certification. Registrants must take and pass the post-test in order to claim SAMs credit(s). SAM credits are offered for the fall meeting and perinatal symposia only.