Editorial
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Preface : Invited Issue Editor, Professor Tai-Tong Wong and the Cancer Predisposition Syndrome

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The *Journal of Korean Neurosurgical Society* (*JKNS*) has published an annual *Pediatric Issue* since 2015, covering various topics in pediatric neurosurgery. Over the past decade (2015–2024), 10 issues have been dedicated to different subjects. For the 2025 *Pediatric Issue*, the selected theme is cancer predisposition syndromes (CPSs). This issue includes 11 articles discussing genetic and clinical aspects of neurofibromatosis (NF) type 1, NF type 2, schwannomatosis, constitutional mismatch repair deficiency (CMMRD) syndrome, Li-Fraumeni syndrome, rhabdoid predisposition syndrome, tuberous sclerosis, von-Hippel Lindau disease, and germline variants in pediatric cancers.

CPSs arise from pathogenic variants in tumor suppressor genes, proto-oncogenes, or DNA repair genes, often with tissue-specific implications. These genetic alterations may occur as germline, constitutional, or mosaic mutations. Approximately 7–10% of all pediatric cancers are associated with hereditary CPSs[14]. In central nervous system tumors, the prevalence is even higher in specific subtypes, such as sonic hedgehog-activated medulloblastoma, atypical teratoid rhabdoid tumors, and choroid plexus carcinomas[6,8,13].

In childhood cancers, the identification of CPSs and the implementation of relevant genetic counseling and surveillance rely on both phenotypic and molecularly confirmed diagnoses. The diagnostic tools used include phenotypic checklists and molecular genetic testing. Generally, the proposed checklist includes family history, the presence of specific tumors, the occurrence of two or more synchronous or metachronous primary neoplasms, and unique phenotypic features such as cutaneous lesions, congenital anomalies, and distinctive facial characteristics[2,10].

The selection of genetic testing is guided by phenotype-based genetic predisposition, clinical indications, the type of mutation (germline, constitutional, or mosaic), and the required se-

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Fig. 1. Portrait of Professor Tai-Tong Wong.

quencing depth. Genetic testing methods include actionable single-gene testing, phenotype-based exome analysis, pediatric CPS multigene panels, and next generation sequencing (NGS)[1-3].

Constitutional or somatic mosaicism in hereditary CPSs plays a significant role in the presentation of atypical phenotypes, influencing clinical severity, specific features, and age of onset[4,12]. The prevalence of mosaicism varies among patients with different CPSs. However, a substantial number of CPS cases remains undetected. With advancements in NGS technology, particularly its increasing sensitivity in DNA analysis, the diagnosis of mosaicism has significantly improved, allowing for the detection of very low-level variant allele frequencies.

Identification of CPS has significant clinical implications for pediatric cancer patients. First, recognizing an underlying CPS can aid in diagnosing specific cancer types, even before a pathological diagnosis, as CPSs are strongly associated with certain cancers, including rare ones.

Second, the presence of germline variants, particularly in tumor suppressor genes, can influence treatment decisions. Cytotoxic therapies that damage DNA, such as alkylating agents or radiation, may exacerbate genetic vulnerabilities, increasing the risk of secondary malignancies in CPS patients[11]. Therefore, personalized treatment strategies tailored to each patient's genetic profile are recommended[7].

Finally, diagnosing CPS enables guided surveillance and preventive measures for both patients and their families[9]. Early cancer detection, prevention of secondary malignancies, and reduced treatment-related toxicity can significantly improve outcomes[5]. Ultimately, a systematic and comprehensive approach enhances the quality of life for children with cancers, while providing informed and sophisticated family counseling.

The 2025 *Pediatric Issue* was organized by the invited editor, Professor Tai-Tong Wong, a world-renowned leader of pediatric neuro-oncology (Fig. 1).

Professor Tai-Tong Wong is a distinguished senior pediatric neurosurgeon who has made significant contributions to the field. He previously served as the President of the International Society for Pediatric Neurosurgery (2008–2009), the Taiwan Society for Pediatric Neurosurgery (2008–2010), and the Asian-Australasian Society for Pediatric Neurosurgery (2015–2017).

After graduating from the National Defense Medical Center in Taiwan in 1973, he completed his neurosurgical training at Taipei Veterans General Hospital (TVGH) in 1978. In 1980, he pursued further training as a clinical and research fellow at The Hospital for Sick Children, Toronto, under the mentorship of Dr. Harold J. Hoffman, Dr. E. Bruce Hendrick, and Dr. Robin P. Humphrey. Since 1981, he has specialized in pediatric neurosurgery at TVGH and was appointed as a professor in 2008.

Dr. Wong's research focuses on clinical and translational studies related to pediatric brain tumors. He has authored over 230 publications in his field of expertise. Currently, he serves as an attending physician in pediatric neurosurgery and director of the Pediatric Brain Tumor Program at Taipei Medical University Hospital (TMUH). Additionally, he is the director of the Department of Pediatric Neurology and Neurosurgery at the Taipei Neuroscience Institute (TNI), Taipei Medical University (TMU).

For this *Pediatric Issue* of the Korean Society for Pediatric Neurosurgery (KSPN) and the *JKNS* on CPSs, we would like to express our sincere gratitude to all contributing authors of the 2025 *Pediatric Issue*. Their valuable contributions encourage us to revisit the clinical significance of CPSs.

AUTHORS' DECLARATION

Conflicts of interest

No other potential conflict of interest relevant to this article was reported.

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