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CHRONIC NEUTROPENIA OF CHILDHOOD

Rose Mary S Stocks

University of Tennessee Health Science Center, USA

Objective: Chronic neutropenia of childhood (CNC) is a rare disorder in which the absolute neutrophil count is below 1500/L over an extended period of time. The objective of this study is to describe the otolaryngologic manifestations associated with CNC to facilitate diagnosis and treatment of this condition.

Methods & Materials: We performed a retrospective chart review of patients with the diagnosis of CNC between 1970 and 2005 at a tertiary pediatric hematology center. After Institutional Review Board approval, 43 patients were evaluated. The average age at hematologic diagnosis was 49 months (range: 1 month – 15 years with 35% <1 year, 44% 1–10 years, 21% >10 years). A total of 2049 encounters were analyzed from the hospital charts.

Results: Twenty four subjects (56%) presented with recurrent otitis media (ROM), sinusitis or pharyngotonsillitis, while 11% presented with oral mucosal lesions. After diagnosis, otolaryngologic problems persisted, including ROM (81%), viral upper respiratory tract infection (67%), oral ulcers or gingivitis (53%), tonsillitis (39%) and sinusitis (37%) and were more common than other systemic infections. Myringotomy tube placement, endoscopic sinus debridement, adenotonsillectomy or tracheostomy were required in 42% of patients. After G-CSF (granulocyte colony-stimulating factor) became available in the early 1990s, the infection rate markedly decreased. Five deaths occurred (12% mortality) including one due to sepsis from otolaryngologic infection.

Conclusion: The majority of children with CNC had otolaryngologic problems at presentation and these continued after diagnosis. While managing common otolaryngologic infections in children, a high index of suspicion for chronic neutropenia is necessary. An otolaryngologist is frequently one of the first physicians to encounter children with this condition. Awareness of CNC and its management will enhance earlier diagnosis and more effective treatment for these children.

Biography

2nd Edition of International Congress on

Pediatrics

Rose Mary S Stocks is the Residency Program Director for Otolaryngology, Head & Neck Surgery Department at UTHSC. She is a fellowship trained Pediatric Otolaryngologist with a Doctorate in Pharmacy and a long bench research history. She has conducted numerous NIH-funded research endeavors with an emphasis on ototoxicity and its prevention in the administration of chemotherapy in the guinea pig model. She has participated in, and published her experience in extra-uterine, prenatal surgical repair of defects of the head and neck (the EXIT procedure). She has served as Research Mentor to numerous residents over the years and greatly increased the amount of scholarly activity within the department. She has taught courses at a national level for the American Academy of Otolaryngology, Head and Neck Surgery on Down's Syndrome for the Otolaryngologist and Management of the Difficult Pediatric Airway. She teaches first year medical students about examination of the ear, nose, and throat and is a tireless surgical educator in the training program. She is a Member of the LeBonheur Multidisciplinary Cleft Lip and Palate Team and devotes her expertise to the care of these patients and their families.

rstocks@uthsc.edu

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