Guest Editorial

Retinoblastoma: Making a Difference Together

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Mary Elizabeth is an Oncology Clinical Nurse Specialist from Memorial Sloan-Kettering Cancer Center in New York City, New York. She graduated from University of Delaware in 1985, with a Bachelor's degree in nursing and followed in 1989, with a Masters of Nursing from Columbia University specializing in oncology. Mary Elizabeth works as a Clinical Nurse Specialist in Memorial Sloan Kettering's ambulatory medical oncology consultative services. Her role includes modeling professional practice to the nurses of the services for which she has responsibility including mentoring and precepting nurses who are new to a practice, coordinating and evaluating their clinical experiences, and overseeing their development in the oncology nursing specialty and the ambulatory clinical practice role. Mary Elizabeth regularly "walks the walk" filling in and assisting in busy practices; this helps her maintain knowledge of the latest advances in disease management as well as experience firsthand the challenges of the ambulatory nursing role. Over the past few years, she has been practicing within MSK's busy and growing world - renowned retinoblastoma (RB) clinic. Mary Elizabeth has presented on RB nationally and at international conferences in the hopes of bringing this curable disease to the attention of nurses worldwide. Early diagnosis and early access to treatment can save lives, eyes, and vision.

This special issue of APJON shines a spotlight on a curable pediatric cancer called RB. RB is the most common primary eye cancer in children and affects approximately 8000 children worldwide. There are no overall cultural or gender differences; incidence rates follow birth rates and range from about 300 cases in the United States per year to 1000 cases per year in China.[1,2] The majority children (70%–75%) present with unilateral disease.[3] All patients with bilateral RB and about 10%–15% of children with unilateral disease carry a germline (hereditary) mutation which is transmissible to their offspring.[4] Children with the germline defect are at higher risk for secondary malignancies, including sarcoma and melanoma.[5,6]

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RB can be cured with a variety of therapies, especially when detected early, and the disease is limited to the eye. Many modalities (and often in combination) are used, including laser photocoagulation, cryotherapy, chemotherapy, radiation, and removal of the eye (enucleation). Enucleation remains a highly effective therapy, often curative alone, and has been the most common treatment for RB[7] historically. Unfortunately, despite the survival rates exceeding 95% in developed countries,[8] worldwide survival rates are dismal with estimated survival for children in low-income countries at 40%.[9] Survival rates are closely related to the stage of disease at presentation; delayed presentation has been

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related to extraocular disease (disease that has migrated outside the eye) and the risk of poor outcome.[1]

Nurses and other health-care clinicians have the power to impact patient outcomes by educating parents and colleagues, implementing screening programs to detect RB in earlier stages, promoting recognition of the curability of this disease, and helping break down barriers to effective treatment. In 2007, Leander *et al*. underwent a large multi-organizational collaborative effort to educate and inform health-care providers in Honduras, about RB. They targeted parents of infants and toddlers participating in an annual vaccination program. Flyers and posters were distributed that explained the severity of RB and the most common presenting sign of leukocoria (a white glow in the eye, also known as "cats eye reflex"). They were able to demonstrate that the addition of a simple, inexpensive educational program to an existing vaccination program improved diagnosis of early stage disease; before the program, 73% of patients were diagnosed with extraocular disease improving to 35% after the implementation.[10]

The concept of including RB screening and education into existing programs and practices is noteworthy. All pediatric nurses, nurse practitioners, and primary care clinicians can increase awareness and education and screen patients at childhood healthcare encounters. The red light reflex is a simple assessment in which a clinician views the retina with an ophthalmoscope expecting symmetric findings bilaterally. The American Academy of Pediatrics recommends performing the red light reflex before discharge from neonatal nursery after birth and at all subsequent routine healthy visits.[11] If there is a family history of RB, screening should be performed following a schedule. Studies have shown that early diagnosis is associated with less advanced disease, allows for local intraocular treatment of small tumors and provides a much better chance of globe preservation.[4,12]

Despite these well-established screening guidelines, Abramson and associates[13] retrospectively studied over 1630 patients with RB at their center and found only 8% of disease was detected by physicians; the majority were detected by family. Parents can detect leukocoria in photographs, especially when the "red eye reduction" function is off; Freeman and Meyer[14] note it would not be too idealistic to suggest that pediatric clinicians use this humble camera technique and save images to the patient's file at each visit.

Unfortunately, screening and early diagnosis are not the only issues concerning disease curability. Despite the increased rate of early diagnosis in Leander *et al*. study above, they described that once diagnosed, one-third of parents refused or abandoned treatment.[10] Abandonment of treatment has been described as much a socioeconomic issue as a medical one, and often various factors beyond the control of the patient and parents are present.[15] Outcomes of parent who abandon treatment for their children have been reported as uniformly dismal.[16]

In the first feature manuscript within this special issue on Retinoblastoma, Biemba K. Maliti describes the current state of RB management in Zambia, Africa. She describes issues with chemotherapy drug availability and equipment maintenance along with a knowledge deficit among primary health-care providers who may prescribe antibiotic eye drops to treat RB, contributing to delayed diagnosis. Maliti reports most children present with advanced disease including 30% with central nervous system and skeletal involvement. She also repeats the theme of treatment abandonment and discusses a study[17] where <10% of children completed planned treatment for RB.

Chawla, Lumar, and Singh examine the influence of socioeconomic and cultural factors on RB management in India in our second featured manuscript. They describe ways in which socioeconomic status affects treatment and prognosis, and they explore the cultural beliefs that influence parental decisions. In this editorial, the authors review some steps to overcome barriers to adequate treatment and list what more can be done to promote positive outcomes. They report their dedicated nursing and social work staff have decreased treatment dropout rates by counseling, educating and tracking RB children and families.

Domingo, Toledo, and Mante also explore factors that influenced parental decisions; they studied enucleation in the Philippines in our third manuscript. Using a cross-sectional descriptive approach, psychosocial barriers and facilitating factors for accepting versus refusing enucleation as a treatment for RB were investigated. They discuss the concept of RB as a fatal illness and parental consideration of the "choice for life" increasing acceptance of enucleation. They suggest healthcare workers help break down barriers and address fears and misconceptions.

I recently had the opportunity to explore why many Chinese parents are reluctant to enucleate, even in the presence of curable disease. I visited Shanghai China with Dr. David Abramson, a world-renowned RB expert along with my institution's international center colleagues to celebrate the collaboration between our clinicians and Chinese colleagues in the care of children with RB (with special thanks to the Elmer and Mamdouha El-Sayed Bobst International Fund and the Fund for Ophthalmic Knowledge). We visited RB clinicians at Xinhua hospital, presented cases, discussed RB management and concluded with a celebration for patients and their families.

Next presented is a focused review of neonatal retinoblastoma: RB that occurs within the 1st month of life.

In general, cancer in neonates differs from cancers occurring later in childhood in terms of incidence, anatomical site, histological features, and clinical behavior.[18,19] Kivela and Hadjistilianou report up to 10% of cases of RB worldwide are diagnosed in the neonatal period and that these cases form a subgroup of RB that has certain typical characteristics. One such characteristic is a positive family history of RB; the authors review case series of children with family history of RB, screened in the prenatal stage and after birth. The authors impress that oncology clinicians and other specialists can be instrumental in the education and counseling of hereditary RB survivors about their risk of secondary neoplasms as well as passing risk of disease onto their children. Knowledge of the psychosocial issues related to genetic testing and consideration and appreciation of cultural and religious beliefs is needed. With improvements and increased access and affordability of genetic testing, perinatal and neonatal screening of children with family history will play a larger role in RB practices.[2]

In the last featured manuscript, this author and colleagues present a close look at a specialized therapy for intraocular retinoblastoma. Ophthalmic artery chemosurgery (OAC) is an interventional surgical procedure in which small doses of chemotherapy are given directly into the arteries perfusing the eye. This highly specialized procedure requires a skilled team and care coordination. OAC has been successfully performed in over 45 countries worldwide.[20] This manuscript outlines the nursing care of children receiving OAC in our center in New York.

The oncology nurse is an integral part of multidisciplinary team caring for a child with RB starting with promotion of screening and early detection. Knowledge of the treatment options and the socioeconomic, cultural, and psychosocial factors that influence parental decisions is vital. RB is curable cancer; we need to improve survival rates worldwide. The sharing of knowledge and expertise and collaborations between institutions and countries are just two examples of how, together, we can make a difference.

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