BERRIEN COUNTY CANCER SERVICE NEWSLETTER

www.bccancerservice.org

In honor of the committed service of Olove Colcord, R.N.

THE MISSION OF THE BERRIEN COUNTY CANCER SERVICE:

To provide free skilled home nursing services, equipment, information and supplies at cost for cancer patients and their families in Berrien County.

January 2012 **(269) 429-3281** **VOLUME XXI ISSUE I**

The **BERRIEN COUNTY CANCER SUPPORT GROUP** is a group for patients, family members and care givers. Come share successes, feelings, fears and practical methods of coping with the physical and emotional aspects of living with the diagnosis of cancer.

The Snowman's Resolution

The snowman's hat was crooked
And his nose was out of place
And several of his whiskers
Had fallen from his face.

But the snowman didn't notice
For he was trying to think
of a New Year's resolution
That wouldn't melt or shrink

He thought and planned and pondered
With his little snowball head
Till his eyes began to glisten
And his toes began to spread:

At last he said, "I've got it!"
I'll make a firm resolve
That no matter what the weather
My smile will not dissolve."

Now the snowman acted wisely
And his resolution won,
For his splinter smile was wooden
And it didn't mind the sun

United Way
United Way of Southwest Michigan

Retinoblastoma cancer

Definition of retinoblastoma: Cancer that forms in the tissues of the retina (the light-sensitive layers of nerve tissue at the back of the eye). Retinoblastoma usually occurs in children younger than 5 years. It may be hereditary or nonhereditary (sporadic).

Incidence

Retinoblastoma is a relatively uncommon tumor of childhood that arises in the retina and accounts for about 3% of the cancers occurring in children younger than 15 years. The estimated annual incidence in the United States is approximately 4 per 1 million children younger than 15 years. Although retinoblastoma may occur at any age, it most often occurs in younger children; the annual incidence is 10 to 14 per 1 million in children aged 0 to 4 years. Ninety-five percent of cases are diagnosed before age 5 years and two-thirds of these cases occur before age 2 years. Older age is usually associated with more advanced disease and a poorer prognosis.

Hereditary and Nonhereditary Forms of Retinoblastoma

Retinoblastoma is a tumor that occurs in heritable (25% to 30%) and nonheritable (70% to 75%) forms. Hereditary disease is defined by the presence of a positive family history, multifocal retinoblastoma, or an identified germline mutation of the RB1 gene. This germline mutation may be known in those patients with a positive family history (25%) or may have occurred in utero at the time of conception, in those patients with sporadic disease (75%). Hereditary retinoblastoma may manifest as unilateral or bilateral disease. The penetrance of the mutation (laterality, age at diagnosis, and number of tumors) is probably dependent on concurrent genetic modifiers, such as MDM2. Most patients with unilateral diseases do not have the hereditary form of the disease, whereas all children with bilateral diseases are presumed to have the hereditary form of the disease, even though only 20% have an affected parent. In hereditary retinoblastoma, tumors tend to occur at a younger age than in the nonhereditary form of the disease. Unilateral retinoblastoma in children younger than 1 year should raise concern for the hereditary disease, whereas older children with a unilateral tumor are more likely to have the nonhereditary form of the disease.

Screening

Children with the hereditary form of retinoblastoma may continue to develop new tumors for a few years after diagnosis. For this reason, children with hereditary retinoblastoma who have a normal examination in at least one eye on initial presentation need to be examined frequently for the development of new tumors. It is recommended that they be examined every 2 to 4 months for at least 28 months. Following treatment, patients require careful surveillance until age 5 years. The interval between exams is based on both the age of the child (more frequent visits as the child ages) and the stability of the disease.

The parents and siblings of patients with retinoblastoma should have screening ophthalmic examinations to exclude an unknown familial disease. Siblings should continue to be screened until age 3 to 5 years or until it is confirmed that they do not have a genetic mutation.

Blood and/or tumor samples can be screened to determine if a retinoblastoma patient has a mutation in the RB1 gene. Commercial laboratories are now available to perform this service. Once the patient's genetic mutation has been identified, other family members can be screened directly for the mutation. The RB1 gene is located within the q14 band of chromosome 13. Exon by exon sequencing of the RB1 gene demonstrates germline mutation in 90% of patients with hereditary retinoblastoma. Although a positive finding with current technology confirms susceptibility, a negative finding cannot absolutely rule it out. The multistep assay includes DNA sequencing to identify mutations within coding exons and immediate flanking intronic regions, Southern blot analysis to characterize genomic rearrangements, and transcript analysis to characterize potential splicing mutations buried within introns. This expanded analysis is showing promise in better defining the functional significance of apparently novel mutations in pilot investigations performed at the University of Pennsylvania. Such testing should be performed only at institutions with expertise in RB1 gene mutation analysis. In cases of somatic mosaicism or cytogenetic abnormalities, the mutations may not be easily detected and more exhaustive techniques such as karyotyping, multiplex ligation-dependent probe amplification (MLPA), and fluorescence in situ hybridization (FISH) may be needed.

The absence of detectable RB1 mutations in some patients may suggest that alternative genetic mechanisms may underlie the development of retinoblastoma.

Genetic counseling should be an integral part of the therapy for a patient with retinoblastoma, whether unilateral or bilateral. It is of utmost importance to assist parents in understanding the genetic consequences of each form of retinoblastoma and to estimate risk of disease in family members. Genetic counseling, however, is not always straightforward. Families with retinoblastoma may have a founder mutation with embryonic mutagenesis causing genetic mosaicism of gametes. A significant proportion (10%–18%) of children with retinoblastoma have somatic genetic mosaicism, making the genetic story more complex and contributing to the difficulty of genetic counseling.

Factors Influencing Mortality

The present challenge for those who treat retinoblastoma is to prevent loss of an eye, blindness, and other serious effects of treatment that reduce the life span or the quality of life. With improvements in the diagnosis and management of retinoblastoma over the past several decades, metastatic retinoblastoma is observed less frequently in the United States and other developed nations. As a result, other causes of retinoblastoma-related mortality in the first decade of life, such as trilateral retinoblastoma and second malignant neoplasms, have become significant contributors to retinoblastoma-related mortality. In the United States, before the advent of chemoreduction as a means of treating bilateral (hereditary) disease, trilateral retinoblastoma contributed to more than 50% of retinoblastoma-related mortality in the first decade after diagnosis.

Trilateral retinoblastoma

Trilateral retinoblastoma is a well-recognized syndrome that occurs in 5% to 15% of patients with hereditary retinoblastoma and is defined by the development of an intracranial midline neuroblastic tumor, which typically develops more than 20 months after the diagnosis of retinoblastoma. Patients who are asymptomatic at the time of diagnosis with an intracranial tumor have a better outcome than patients who are symptomatic.

Given the poor prognosis of trilateral retinoblastoma and the short interval between the diagnosis of retinoblastoma and the occurrence of trilateral disease, routine neuroimaging could potentially detect the majority of cases within 2 years of first diagnosis. While it is not clear whether early diagnosis can impact survival, the frequency of screening with magnetic resonance imaging (MRI) for those suspected of having hereditary disease or those with unilateral disease and a positive family history has been recommended as often as every 6 months for 5 years. It is unclear if this will have an impact on outcome or survival. Computed tomography scans should be avoided for routine screening in these children because of the perceived risk of exposure to ionizing radiation.

Second malignant neoplasms

Patients with hereditary retinoblastoma have a markedly increased frequency of second malignant neoplasms (SMN). The cumulative incidence was reported to be 26% (± 10%) in nonirradiated patients and 58% (± 10%) in irradiated patients by 50 years after diagnosis of retinoblastoma—a rate of about 1% per year. However, more recent studies have reported the rates to be about 9.4% in nonirradiated patients and about 30.4% in irradiated patients. Most of the SMN are osteosarcomas, soft tissue sarcomas, or melanomas. There is no evidence of an increased incidence of acute myeloid leukemia in children with hereditary retinoblastoma.

A cohort study of 963 patients, who were at least 1-year survivors of hereditary retinoblastoma diagnosed at two U.S. institutions from 1914 through 1984, evaluated risk for soft tissue sarcoma overall and by histologic subtype. Leiomyosarcoma was the most frequent subtype, with 78% being diagnosed 30 or more years after the retinoblastoma diagnosis. Risks were elevated in patients treated with or without radiation therapy, and, in those treated with radiation therapy, sarcomas were seen both within and outside the field of radiation. The carcinogenic effect of radiation increased with dose, particularly for secondary sarcomas where a step-wise increase is apparent at all dose categories. In irradiated patients, two-thirds of the second cancers occur within irradiated tissue and one-third occur outside the radiation field.

The risk for SMN is heavily dependent on the patient's age at the time the external-beam radiation therapy is given, especially in children younger than 12 months, and the histopathologic type of SMN may be influenced by age. These data support a genetic predisposition to soft tissue sarcoma, in addition to the risk of osteosarcoma.

It has become apparent that patients with hereditary retinoblastoma are also at risk of developing epithelial cancers late in adulthood. A marked increase in mortality from lung, bladder, and other epithelial cancers has been described.

Survival from second malignancies is certainly suboptimal and varies widely across studies. However, with advances in therapy, it is essential that all second malignancies be treated with curative intent. Those who survive SMN are at a 7-fold increased risk for developing a subsequent malignancy. The risk further increases 3-fold when patients are treated with radiation therapy for their retinoblastoma. There is no clear increase in second malignancies in patients with sporadic retinoblastoma beyond that associated with the treatment.

Late Effects from Retinoblastoma Therapy

Patients with retinoblastoma demonstrate a variety of long-term visual field defects after treatment for their intraocular disease. These defects are related to tumor size, location, and treatment method. One study of visual acuity following treatment with systemic chemotherapy and focal ophthalmic therapy was conducted in 54 eyes in 40 children. After a mean follow-up of 68 months, 27 eyes (50%) had a final visual acuity of 20/40 or better, and 36 eyes (67%) had final visual acuity of 20/200 or better. The clinical factors that predicted visual acuity of 20/40 or better were a tumor margin at least 3 mm from the foveola and optic disc and an absence of subretinal fluid.

Since systemic carboplatin is now commonly used in the treatment of retinoblastoma (Refer to Intraocular Retinoblastoma and Extraocular Retinoblastoma sections of this summary), concern has been raised about hearing loss related to therapy. However, an analysis of 164 children treated with six cycles of carboplatin-containing therapy (18.6 mg/kg per cycle) showed no loss of hearing among children who had a normal initial audiogram.

Cellular Classification

Retinoblastoma is composed mainly of undifferentiated anaplastic cells that arise from the retina. Histology shows similarity to neuroblastoma and medulloblastoma, including aggregation around blood vessels, necrosis, calcification, and Flexner-Wintersteiner rosettes. Retinoblastomas are characterized by marked cell proliferation as evidenced by high mitosis counts and extremely high MIB-1 labeling indices

Although there are several staging systems available for retinoblastoma, for the purpose of treatment, retinoblastoma is categorized into intraocular and extraocular disease.

Intraocular

5-year disease-free survival: >90%

Intraocular retinoblastoma is localized to the eye and may be confined to the retina or may extend to involve other structures such as the choroid, ciliary body, anterior chamber, and optic nerve head. Intraocular retinoblastoma, however, does not extend beyond the eye into the tissues around the eye or to other parts of the body.

Extraocular

5-year disease-free survival: <10%

Extraocular (metastatic) retinoblastoma has extended beyond the eye. It may be confined to the tissues around the eye, or it may have spread to the central nervous system, bone marrow, or lymph nodes.

Reese-Ellsworth Classification for Intraocular Tumors

Reese and Ellsworth developed a classification system for intraocular retinoblastoma that has been shown to have prognostic significance for maintenance of sight and control of local disease at a time when surgery and external-beam radiation therapy (EBRT) were the primary treatment options.

Group I: very favorable for maintenance of sight

1. Solitary tumor, smaller than 4 disc diameters (DD), at or behind the equator.
2. Multiple tumors, none larger than 4 DD, all at or behind the equator.

Group II: favorable for maintenance of sight

1. Solitary tumor, 4 to 10 DD at or behind the equator.
2. Multiple tumors, 4 to 10 DD behind the equator.

Group III: possible for maintenance of sight

1. Any lesion anterior to the equator.
2. Solitary tumor, larger than 10 DD behind the equator.

Group IV: unfavorable for maintenance of sight

1. Multiple tumors, some larger than 10 DD.
2. Any lesion extending anteriorly to the ora serrata.

Group V: very unfavorable for maintenance of sight

1. Massive tumors involving more than one half of the retina.
2. Vitreous seeding.

International Classification System for Intraocular Retinoblastoma

There is a new classification system for retinoblastoma, which may offer greater precision in stratifying risk for newer therapies. The International Classification for Intraocular Retinoblastoma that is used in the current Children's Oncology Group treatment studies, as well in some institutional studies, has been shown to assist in predicting those who are likely to be cured without the need for enucleation or EBRT.

- Group A: Small intraretinal tumors away from foveola and disc.
    - All tumors are 3 mm or smaller in greatest dimension, confined to the retina and
    - All tumors are located further than 3 mm from the foveola and 1.5 mm from the optic disc.
- Group B: All remaining discrete tumors confined to the retina.
    - All other tumors confined to the retina not in Group A.
    - Tumor-associated subretinal fluid less than 3 mm from the tumor with no subretinal seeding.
- Group C: Discrete local disease with minimal subretinal or vitreous seeding.

    Tumor(s) are discrete.
    Subretinal fluid, present or past, without seeding involving up to one-fourth of the retina.
    Local fine vitreous seeding may be present close to discrete tumor.
    Local subretinal seeding less than 3 mm (2 DD) from the tumor.
- Group D: Diffuse disease with significant vitreous or subretinal seeding.
    - Tumor(s) may be massive or diffuse.
    - Subretinal fluid present or past without seeding, involving up to total retinal detachment.
    - Diffuse or massive vitreous disease may include "greasy" seeds or avascular tumor masses.
    - Diffuse subretinal seeding may include subretinal plaques or tumor nodules.
- Group E: Presence of any one or more of the following poor prognosis features.
    - Tumor touching the lens.
    - Tumor anterior to anterior vitreous face involving ciliary body or anterior segment.
    - Diffuse infiltrating retinoblastoma.
    - Neovascular glaucoma.
    - Opaque media from hemorrhage.
    - Tumor necrosis with aseptic orbital cellulites.
    - Phthisis bulbi.

Treatment Option Overview

Treatment planning by a multidisciplinary team of cancer specialists, including a pediatric oncologist, ophthalmologist, and radiation oncologist, who have experience treating ocular tumors of childhood is required to optimize treatment planning.

The goals of therapy are threefold:

1. Eradicate the disease to save the patient's life.
2. Preserve as much vision as possible.
3. Decrease risk of late sequelae from treatment, particularly second malignant neoplasms.

The type of treatment required depends on both the extent of the disease within the eye and whether the disease has spread beyond the eye, either to the brain or to the rest of the body. Eyes with glaucoma and those in which glaucoma resulted in buphthalmia are significantly associated with high-risk pathology risk factors and the occurrence of microscopically residual tumor. Enucleation is reserved for patients with advanced unilateral intraocular disease with no hope for useful vision in the affected eye. Subsequent risk of extraocular recurrence may be increased in the presence of high-risk histopathologic features such as massive choroid invasion, scleral invasion, and optic nerve invasion. Clinical features predictive of these histological findings include eyes with glaucoma, especially those that have become buphthalmic. Routine bone marrow biopsy and lumbar puncture are not indicated, except when there is a high level of suspicion that the tumor has spread beyond the globe. Examples include patients with an abnormal complete blood count or those whose tumors show massive choroidal involvement and which extend beyond the lamina cribrosa on pathologic examination of the enucleated specimen.

It is not uncommon for patients with retinoblastoma to have extensive disease within one eye at diagnosis, with either massive tumors involving more than one-half of the retina, multiple tumors diffusely involving the retina, or obvious seeding of the vitreous. For those with bilateral disease, systemic therapy may be used to treat the more severe eye. There are data suggesting that the use of systemic chemotherapy may decrease the risk of development of trilateral retinoblastoma.

BCCS donations to Berrien Community Foundation

During the time period of January 1- December 31, 2012, the Berrien Community Foundation will provide a match of .50 cents (up to a maximum of $1,000) on each dollar contributed to the Berrien County Cancer Service Endowment Fund at the Berrien Community Foundation. Checks should be made out to BCF/Berrien County Cancer Service Endowment Fund and mailed directly to the Foundation at the address below:

Berrien Community Foundation
2900 South State Street, Suite 2 East
St. Joseph, MI 49085

BCCS Famous Spiced Almonds

1 pound of whole almonds
1 cup sugar
1 teaspoon salt
1 teaspoon cinnamon
1 tablespoon water
1 egg white

Beat egg white with water until frothy. Add nuts and coat thoroughly. Mix with whisk sugar, cinnamon and salt.

Coat almonds with sugar mixture. Place almonds on cookie sheet sprayed with Pam. Bake at 300° for 40-45 minutes stirring every 15 minutes until golden brown being careful not to scorch.

Chicken with Cashews and Snow Peas

1 whole chicken breast, split, skinned, boned and cut into 1 inch cubes
2 cloves garlic, peeled and minced
1 tablespoon soy sauce
1 tablespoon dry sherry
2 tablespoon cornstarch
1 teaspoon hoisin sauce
1 tablespoon peanut oil
20 now peas, ends and strings removed
½ cup sliced water chestnuts, drained
½ cup hot chicken stock
½ teaspoon salt
½ cup unsalted raw cashews

Marinate chicken 15 minutes in mixture of next 5 ingredients. Heat oil in uncovered wok at 375°. Add chicken mixture; stir-fry 3 minutes. Add snow peas and water chestnuts; stir-fry 30 seconds. Add stock and salt; stir-fry until slightly thickened. Stir in cashews. Serve immediately.

In Loving Memory

During November 2011, Memorial Donations were generously made by and for the following people

In Memory of Henry Angelo
Leonard & Dorothy Krumrie, St. Joseph

In Memory of Allen Henry Cable
Brett & Jennifer Costanza, Sodus

In Memory of John "Jack" Erwin
Linda Knoll, New Buffalo
% Your Friends at Exit 1 Realty
Jill Morowitz, Chicago IL
Vito & Frances Rizzi, Park Ridge IL
Joe & Marilyn Thompson, Connersville IN

In Memory of Edward Fester
Harriet Fester, Bridgman

In Memory Terry Hendrickson
Paul & Phyllis Huss, Howell

In Memory of Joseph Gene Kamp
Darrell & Betty Gast, Baroda

In Memory of Mary Ann Mashke
Nancy Green, Benton Harbor

In Memory of Debra Silverthorn
Virginia Antonson, St. Joseph
Patricia E. Greenburg, Stevensville
Dick & Loretta Kasmer, St. Joseph
Linda Korrell, Benton Harbor
Molly M. Moran, St. Joseph
John H. Reed, Benton Harbor
Camille Sandel, Stevensville
St. Joseph Public Schools Faculty & Staff, St. Joseph
Ray & Kay Van Patten, Bridgman
Mike & Tracy Wagner, St. Joseph

In Memory Of Kenneth Smith
Elaine M. White, Watervliet

In Memory of Louis B. Vogl
Rick & Suzanne Birkhold, Dowagiac
Mr. & Mrs. G. D. Claussen, Menomonee Falls WI
Jennifer Gray, Niles

In Memory of Michael Welihan
Ann L. Welihan, St. Joseph

Berrien County Cancer Service sends our sincere sympathy to all those who have recently lost loved ones. We thank all of our generous donors. Your donations are very much appreciated and will help cancer patients in Berrien County. Thank you.

Looking Ahead

BCCS SUPPORT GROUP – Stevensville
January 3 & 17 – 1:30 p.m.
February 7 & 21 – 1:30 p.m.

BCCS SUPPORT GROUP – Niles
January 10 & 24 – 1:30 p.m.
February 14 & 28 – 1:30 p.m.

OSTOMY SUPPORT GROUP – Stevensville
January 17 – 1:30 p.m.
February 21 – 1:30 p.m.

RAINBOWS OF HOPE– St. Joseph
Marie Yeager Cancer Center
January 12 – 5:30 p.m.
February 9 – 5:30 p.m.

OSTOMY SUPPORT GROUP
Lakeland Regional Medical Center
January 12 – 6:00 p.m.
February 9 – 6:00 p.m.

MAN TO MAN
PROSTATE SUPPORT GROUP
Trinity Center, St. Joseph
January 17 – 6:30 p.m.
February 21 - 6:30 p.m.

DATES TO REMEMBER IN JANUARY
January 1 – New Years Day
January 7 – I'm not going to take it anymore Day
January 10 – United Nations Day
January 13 – Friday the 13th
January 15 - Humanitarian Day
January 21 – National Hugging Day
January 24 – Belly Laugh Day
January 27 – Fun at Work Day

Newsletters available online
Our newsletters are available on our website: *www.bccancerservice.org*. If you would like to be removed from this mailing list, please call our office at 269-429-3281 or send us an e-mail: *staff@bccancerservice.org*.

Thank you!

Please Consider...

Berrien County Cancer Service, Inc., is a non-profit organization funded primarily by the United Way, private donations and fund-raisers. We receive no Medicare, Medicaid or other insurance payments. To continue our free services to Berrien County cancer patients, we need your help. Any donation is greatly appreciated.

Donations to our General Fund will help balance our current budget. Donations to our Endowment Fund will help guarantee that the Cancer Service will be available for as long as needed. Your contribution to our non-profit 501(c)(3) corporation is tax deductible – an acknowledgment and receipt for tax purposes will be sent.

Donations can be made in honor of someone or in memory of a loved one. In these instances, we would also like to send acknowledgment to the honoree or next-of-kin so please provide that information when making your donation.

_____ General Fund _____Endowment Fund

Your Name _____

Your Address _____

City/State/Zip_____

_____

Donation Amount $ _____

_____

OR

In Memory of _____

Please send notification of my gift to:

Name_____

Address_____

City/State/Zip_____

Thank you for your generosity!

Berrien County Cancer Service, Inc.
7301 Red Arrow Highway
Stevensville, MI 49127

RETURN SERVICE REQUESTED

NONPROFIT ORGANIZATION
U.S. POSTAGE PAID
STEVENSVILLE, MI
PERMIT NO. 129

CANCER SUPPORT GROUP – Stevensville Office
1st and 3rd Tuesday of each month - 1:30 p.m.
Berrien County Cancer Service, Inc.
7301 Red Arrow Highway
Stevensville, MI 49127
Phone: (269) 429-3281 or (269) 465-5257

CANCER SUPPORT GROUP – Niles
2nd and 4th Tuesday of each month – 1:30 p.m.
Niles Senior Center
1109 Bell Road
Niles, MI 49120
Phone: (269) 429-3281

RAINBOWS OF HOPE GROUP- St. Joseph
2nd Thursday of each month – 5:30 p.m.
Marie Yeager Cancer Center
Ward and Kinney Room
3900 Hollywood Rd.
St. Joseph, MI 49085
Phone: (269) 556-7114

OSTOMY SUPPORT GROUP
2nd Thursday of each month- 6:00 p.m.
Lakeland Regional Medical Center
Community Room
1234 Napier Ave
St. Joseph, MI 49085
Phone: (269) 983-8804

MAN TO MAN – Prostate Support Group
3rd Tuesday of each month – 6:30 p.m.
Trinity Center
619 Main Street (use Main entrance)
St. Joseph, MI 49085
Phone: (800) 465-5244

Printing Courtesy of Lakeland Community Benefit