Practice Guidelines for Patients with Rare Cancers

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Although rare cancers affect few individuals, those who cope with them experience the range of emotions brought on by a cancer diagnosis as well as the difficulties associated with unusual diseases. The detection and identification of rare cancers can require extended workup, costly travel for consultation, and investment in relationships with multiple treatment teams. When a symptom profile matches no well-known syndrome, patients may encounter considerable skepticism, hear their symptoms described as psychosomatic, or have to insist to complete diagnostic testing. Patients who receive conflicting medical opinions must initiate treatment in the absence of consensus; data supporting a recommended treatment may derive from just 5 or 10 cases. Some families agonize about making the wrong decision, delaying treatment. Often, the combined effect of these experiences is heightened anxiety. Even though well trained to intervene in emotionally charged situations, oncology professionals themselves lack information about the psychosocial needs of patients and families isolated by the rarest of circumstances.

About Rare Cancers

Although there appears to be no generally accepted definition among oncology professionals for what constitutes a rare cancer, the Orphan Drug Act[1] designates an orphan disease as one that strikes fewer than 200,000 persons in the United States. Approximately 5000 rare disorders affect 20 million Americans, many of whom spend 6 years or more seeking a diagnosis.[2] Examples of cancers and related conditions that are identified in medical journals as rare include rare primary cancers[3]; uncommon sites of metastasis[4]; cancers with unusual presenting symptoms,[5] manifestations,[6] or complications[7]; and cancers that rarely affect a particular age group[8] or population.[9] The American Cancer Society categorizes all childhood cancers as rare.[10] Patient information is relatively scarce for sites not typically profiled among the 10 or 12 most common cancers.[10–12]

Information Hunger and Overload

Many patients ask for and receive disease-specific publications to assist their participation in treatment decisions, an option not available for individuals with rare cancers. The lack of accessible educational material geared to the lay public can greatly compound the "normal" or anticipated anxiety of a new cancer diagnosis, particularly for those patients who confront stress by gathering information. In the absence of such resources, medical consumers often tackle professional journals in the hope of finding any reference to their condition. Similarly, health organizations share excerpts from medical texts with clients who have rare cancers. This popular strategy fails to address the needs of persons with low literacy skills.[13] Moreover, not everyone realizes that these data were not meant to be applied literally to individual circumstances. Technical literature may dwell on worst cases, long-term complications, or may

Table 1. ***Rare Cancers: Guidelines for Information Gathering***

- Be aware that some Internet sites and agencies charge fees for search-related services.
- Encourage patients to involve medical librarians or resource room volunteers.
- Discuss the technical nature of available materials.
- If nothing is found, check for word reversals or embedded names.
- If this is a common tumor rarely detected at this site, widen search parameters; contact the clinic with the greater experience.
- If the diagnosis is a new classification, be aware that little information may be available.
- Use Interlibrary Loan to access locally unavailable journals.
- For non-English articles, seek a patient education department for translation.
- Look for recurring names in references; search for literature reviews.
- Independently verify treatment and referral information obtained online.
- Try a large database such as the National Institute Cancer Information Service (1-800-4-CANCER); look to cancer centers and pharmaceutical companies.
- Consider networking with other patients, even if the diagnosis is not an exact match.

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Queries and contributions for Patient Resources may be sent to the Department Editors at *Cancer Practice*, American Cancer Society, 1180 Avenue of the Americas, New York, NY 10036.

Table 2. *Support Organizations and Internet Resources*

Rare Disorders or Rare Cancers: General

National Organization for Rare Disorders
P.O. Box 8923
New Fairfield, CT 06812-8923
(800) 999-6673 * (203) 746-6518
FAX: (203) 746-6481
E-mail: orphan@nord-rdb.com
http://www.NORD-RDB.com/~orphan
Coalition of more than 100 rare disease agencies. Rare disease database (includes several cancers), physician database, support, and advocacy.

MUMS National Parents Network (Mothers United for Moral Support)
150 Custer Court
Green Bay, WI 54301
(920) 336-5333
E-mail: mums@netnet.net
www.waisman.wisc.edu/~rowley/mums/home.htmlx
International support and referral network for families of children with rare conditions.

Best Doctors Care Access Network
1359 Silver Bluff Road
Aiken, SC 29803
(888) DOCTORS (1-888-362-8677)
E-mail: info@bestdoctors.com
http://www.bestdoctors.com
Fee-based referral network designed for the treatment of rare, complex, and chronic "high-acuity" cases requiring uniquely skilled physicians.

Alliance of Genetic Support Groups
4301 Connecticut Avenue NW, Suite 404
Washington, DC 20008
(800) 336-4363 * (202) 966-5557
E-mail: medallianc@aol.com
http://medhlp.netusa.net/www/agsg.htm
A nonprofit referral organization for persons coping with genetic disorders.

NIH Office of Rare Diseases
National Institutes of Health
31 Center Drive, MSC-2082
Room 1B-03
Bethesda, MD 20892-2082
(301) 402-4336
E-mail: sg18b@nih.gov
http://rarediseases.info.nih.gov/ord
Information about more than 6000 rare diseases, clinical trials, current research, and support groups.

National Parent to Parent Support and Information System, Inc.
P.O. Box 907
Blue Ridge, GA 30513
(800) 651-1151 * (706) 632-8822
E-mail: judd103w@wonder.em.cdc.gov
http://www.nppsis.org
Federally funded program linking parents of children with rare disorders; provides referrals.

One in a Million Kids
P.O. Box 156
Seal Beach, CA 90740
(310) 588-2562
E-mail: ezrasam@aol.com
Support and resources for parents/care givers of children with rare or undiagnosed disorders. Disability awareness program available.

American Self-Help Clearinghouse
Northwest Covenant Medical Center
25 Pocono Road
Denville, NJ 07834-2995
(973) 625-9565
E-mail: ASHC@cybernex.net
http://www.cmhc.com/selfhelp
Offers a listing of hard-to-find self-help groups.

Specific Tumor Sites Not Listed Among Leading Cancers

American Brain Tumor Association
2720 River Road, Suite 146
Des Plains, IL 60018
(800) 886-2282 * (847) 827-9910
E-mail: info@abta.org
http://www.abta.org

International Myeloma Foundation
2129 Stanley Hills Drive
Los Angeles, CA 90046
(800) 452-CURE
E-mail: TheIMF@aol.com
http://www.myeloma.org

American Liver Foundation (Liver cancer)
1425 Pompton Avenue
Cedar Grove, NJ 07009
(800) 223-0179 * (201) 256-2550
E-mail: info@liverfoundation.org
http://www.liverfoundation.org

National Association for Parents of the Visually Impaired (retinoblastoma)
P.O. Box 317
Watertown, MA 02272
(800) 562-6265
No current E-mail address or web site.

Table 2. ***Continued***

Specific Tumor Sites Not Listed Among Leading Cancers (continued)

National Brain Tumor Foundation
785 Market Street, Suite 1600
San Francisco, CA 94103
(800) 934-CURE
E-mail: nbtf@braintumor.org
http://www.braintumor.org

Neuroblastoma Children's Cancer Society
P.O. Box 957672
Hoffman Estates, IL 60195
(800) 532-5162
(847) 490-4240
http://www.granitewebworks.com/nccs.htm

The Carcinoid Cancer Foundation, Inc.
1751 York Avenue
New York, NY 10128
(212) 722-3132
E-mail: info@carcinoid.org
http://www.carcinoid.org

National Carcinoid Support Group, Inc.
P.O. Box 44233
Madison, WI 53744-4233
E-mail: jean@mick.com
http://members.aol.com/thencsg/index.html

The Brain Tumor Society
84 Seattle Street
Boston, MA 02134
(800) 770-8287 * (617) 783-0340
E-mail: info@tbts.org
http://www.tbts.org

VHL Family Alliance (von Hippel-Lindau)
171 Clinton Road
Brookline, MA 02146
(800) 767-4845 * (617) 232-5946
E-mail: info@vhl.org
http://www.vhl.org

Additional Internet Resources

Steve Dunn's Cancer Guide: Some Advice on Dealing with a Rare Cancer
http://cancerguide.org/rare_cancer.html

Online support groups for rare cancers
Available through OncoLink's automated e-mail discussion group subscriber:
RARE-CANCER, AA-MDS-TALK (aplastic anemia and myelodysplastic syndrome), ADEN-CYST (adenoid cystic carcinomas), BRAINTMR, CARCINOID, CTCL-MF (cutaneous T-cell lymphoma-mycosis fungoides), DESMOID, EC-GROUP (esophageal cancer); E-Sarcoma (Ewing sarcoma), GVHD (graft_versus_host disease), IBC (inflammatory breast cancer), KAPOSI, LARYNX-C, L-M-SARCOMA (leiomyosarcoma), LIVER-ONC, LYMPHEDEMA, MaleBC (male breast cancer), Mantlecell (mantle cell lymphoma), MYELOMA, PANCREAS-ONC, R-BLASTOMA, SARCOMA, STOMACH-ONC, TC-NET (testicular cancer), THYROID-ONC, WSMG-NET (Waldenstrom's macroglobulinemia cancer), as well as several discussion groups for more common cancers.

Sarcoma Central
http://www.charm.net/~kkdk/sarcoma_html

OncoLink
http://cancer.med.upenn.edu
http://cancer.med.upenn.edu/forms/listserv.html.
See cases of the month as well as the automated e-mail discussion group subscriber.

cancerONLINE (includes a section on less common adult cancers)
http://www.stonecottage.com/canceronline

Rare Genetic Diseases in Children: An Internet Resource Gateway
http://mcrcr2.med.nyu.edu/murphp01/homenew.htm

bluntly evaluate prognostic factors as a means of educating physicians. Adaptive denial[14] is difficult to maintain because extended information searches can create distress at a time when families are seeking reassurance and hope.

Oncology professionals, who rely increasingly on written materials to reinforce patient teaching, may balk at the disproportionate time and effort required to locate resources for this population. Guidelines that can direct efforts to gather information are listed in Table 1. When they exist at all, organizations for persons with rare diagnoses (Table 2) seem particularly inaccessible as they may lack toll-free numbers, relocate often, or operate out of a patient's or parent's home office. Professionals find that they must verify and update such databases frequently.

Building Support Networks

Without someone to mirror (and therefore, normalize) their experience, persons with rare cancers may more

readily reach out to others, even across great distances, using programs that link participants by diagnosis. Most patients insist on an exact match, as if someone only with the same cancer cell type could truly understand their situation, while declining a psychosocial match based on common demographics. As an interim measure, oncology professionals can encourage patients to consider networking with someone who has a different but also uncommon cancer. This strategy reduces isolation while patients continue to search for a closer match. Professionals can also prepare patients for the possibility that their rare illness may manifest differently—for better or for worse—in another individual and that matching by level of disability is another option.

Another strategy to counteract isolation is to access e-mail discussion groups. A growing number of these networks (Table 2) serves persons with rare cancers. Reviewing the potential strengths and weaknesses of such programs may help consumers make an informed choice about participation. To minimize pitfalls, practitioners may encourage patients to protect themselves by inquiring how telephone volunteers or listserv administrators are recruited and trained.

Telling One's Story: Assessing Readiness

Patients may wish to tell their story to help them think through their decisions, promote healing, offer future patients information and reassurance, educate healthcare professionals, or celebrate survivorship and express gratitude to the medical community. Writing one's story can empower the storyteller; it can also greatly increase feelings of vulnerability and exposure. Although some persons with rare cancers eventually do tell their story, practitioners should avoid conveying the message, however indirectly, that this is a compulsory part of healing. For example, several popular reference and self-help books have been written by persons coping with rare illnesses; in sharing these materials, practitioners should mention that not all persons cope in the same manner.

Conclusion

Because patient education materials are scarce for persons coping with rare cancers, patients who seek information must rely heavily on medical journals and other materials written for healthcare professionals. As a supplement, patient educators can prepare a handout that outlines common psychosocial concerns expressed by those with rare cancers, regardless of site, and which includes general resource listings. For the emotional resonance[14] absent in technical writing, practitioners can selectively recommend accounts by survivors of rare disorders written for newsletters, magazines, or books, even if the diagnosis is not an exact match.

References

1. Pub L No. 97-414 (FDA); 21 USC 360ee.
2. National Organization for Rare Disorders. *Out of the Darkness, into the Light.* Fairfield, Conn: National Organization for Rare Disorders; 1996:2.
3. Mordkin RM, Dahut WL, Lynch JH. Renal angiosarcoma: a rare primary genitourinary malignancy. *South Med J.* 1997;90(11):1159–1160.
4. Williams HK, Brown AM, Barber PC. Parotid gland metastases of two uncommon neoplasms with low metastatic potential. *J Oral Pathol Med.* 1997;26(8):388–392.
5. Panos A, Kalangos A, Sztajzel J. Left atrial myxoma presenting with myocardial infarction: case report and review of the literature. *Int J Cardiol.* 1997;62(1):73–75.
6. Campbell DC, Grippaudo FR, Lewandowski R. Congenital leukemia cutis: an unusual manifestation of a rare disease. *Plast Reconstr Surg.* 1997;100(7):1809–1811.
7. Anderson AS, Krauss D, Lang R. Cardiovascular complications of malignant carcinoid disease. *Am Heart J.* 1997;134(4):693–702.
8. Orditura M, De Vita F, Catalano G. Adult Wilms' tumor: a case report. *Cancer.* 1997;80(10):1961–1965.
9. Ceresoli GL, Ferreri AJ, Bucci E, Ripa C, Ponzoni M, Villa E. Primary cardiac lymphoma in immunocompetent patients: diagnostic and therapeutic management. *Cancer.* 1997;80(8):1497–1506.
10. American Cancer Society. *Cancer Facts and Figures.* Atlanta; Ga: American Cancer Society; 1998:9.
11. Fact sheet: twelve major cancers. *Sci Am.* 1996;275(3):126–132.
12. Morra M, Potts E. Dictionary of unusual cancers. *Choices.* New York: Avon Books; 1994:710–714.
13. Morra ME, Varricchio C. Teaching patients with limited reading skills. *Cancer Practice.* 1993;1(2):154–156.
14. Dreifuss-Kattan, E. *Cancer Stories: Creativity and Self-Repair.* Hillsdale, NJ: Analytic Press; 1990.