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Aller au sommaire du numéro

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Résumé de l'article

Dans cet article, je décris quelques-unes des raisons pour lesquelles les « previvors » de BRCA (c-a-d. « sur**viv**ants d'une **pré**disposition au cancer ») sont différents des previvors avec d'autres cancers héréditaires. J'examine comment l'absence d'une norme de soins pour le risque de cancer du sein chez les femmes ayant une mutation BRCA, associée à un large éventail de pénétration génétique et une mortalité plus faible, rend le BRCA différent des autres cancers héréditaires qui ont des directives claires et établies. En plus de ces différences médicales, des facteurs sociaux tels que la prééminence culturelle du cancer du sein et la signification sociale des seins ont engendré une identité prédictive individuelle plus complexe et une réponse culturelle aux femmes ayant une mutation BRCA.

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Mots clés
previvor, BRCA, cancer héréditaire, cancer du sein

Summary

In this paper, I outline some of the reasons why BRCA "previvors" (i.e., "sur**vivors** of a **pre**disposition to cancer") are different from previvors with other hereditary cancers. I examine how the absence of a standard of care for breast cancer risk for women with a BRCA mutation, coupled with a broad range of genetic penetrance and lower mortality, makes BRCA different than other hereditary cancers that have clear and established guidelines. In addition to these medical differences, social factors like the cultural prominence of breast cancer and the social significance of breasts have engendered a more complicated individual previvor identity for and cultural response to women with a BRCA mutation.

Keywords
previvor, BRCA, hereditary cancer, breast cancer

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Introduction

Angelina Jolie made headlines when she revealed that she has one of the BRCA genetic mutations,[1] which substantially increases her risk of developing breast and ovarian cancer. The big news was not that she had this mutation; it was her decision to have a prophylactic mastectomy in order to minimize the likelihood of getting breast cancer. The cultural narrative of cancer survivors is ubiquitous. In recent years, there is a new cultural narrative of individuals battling with cancer; however, these individuals do not in fact have cancer nor (in most cases) have they ever had cancer. These individuals, known as "previvors" (short for "sur**vivor** of a **pre**disposition to cancer"), often feel like they too are facing a cancer battle because they face a high risk of developing cancer due to a positive result on a genetic test for hereditary cancer, a family history, or any other predisposing factor. In the medical literature, these individuals are referred to as "unaffected carriers"; yet for many, the term unaffected carrier does not convey the challenges they face and how they understand themselves. The term "previvor" emerged in 2000 on a message board hosted by Facing Our Risk of Cancer Empowered (FORCE), a nonprofit organization dedicated to fighting hereditary breast and ovarian cancer (HBOC), in response to a contributor who wanted a label to describe her cancer fight [1].

Despite the broad definition of previvor, discussions of previvors generally refer to women who have tested positive for one of the BRCA mutations. Yet, there are other hereditary cancers for which prophylactic treatment is available. For example, familial adenomatous polyposis (FAP) is an inherited syndrome associated with an alteration in the MUTYH or APC gene that leads to colon and rectal cancer and standard treatment is a prophylactic colectomy. These other types of hereditary cancers are generally not included in discussions of previvors and the previvor identity is not as common among individuals with these other hereditary cancers. Especially given that BRCA genetic testing was predated by genetic tests for other hereditary cancers [2], why has the previvor identity only recently emerged and stayed within the BRCA community? Comparing BRCA to FAP (see Table 1), I outline some medical and social factors that have played a role in this phenomenon.

Table 1: HBOC versus FAP

Hereditary syndrome	**Hereditary breast and ovarian cancer (HBOC)**	**Familial adenomatous polyposis (FAP)**
Genes mutated	BRCA 1 and BRCA 2	APC and MUTYH
Organ(s) affected	Breast and ovary	Colon
Year gene(s) discovered	1994 and 1995	1991
Genetic test reliability	88%	80-90%
Percent of breast/ovarian and colon cancer that is genetic	5-10%	1%
Malignancy	Breast: 45-85%, Ovary: 11-62%	100%
Average age cancer first develops	30-49	mid-teen
Is prophylactic surgery standard of care?	Breast: no; Ovary: yes	Yes
Average age of prophylactic surgery	Late 20s-40s	18-20
Mortality without prophylactic surgery	Depends, possibility of normal lifespan	100%

Specifically, I examine how the absence of a standard of care for breast cancer risk for women with a BRCA mutation, coupled with a broad range of genetic penetrance and lower mortality, makes BRCA different from FAP and other hereditary cancers that have clear and established guidelines. I focus mainly on breast cancer risk associated with BRCA, and not on ovarian cancer risk, because of the

[1] Although there are medical differences between the BRCA1 and the BRCA2 genetic mutations, for the sake of simplicity, I will be discussing both of them together unless otherwise noted.

lack of guidelines for handling breast cancer risk and because of cultural factors surrounding breasts and breast cancer. In short, the lack of guidelines for breast cancer risk combined with the cultural prominence of breast cancer and the social significance of breasts has engendered a more complicated individual previvor identity and cultural response than for hereditary cancers where there are established guidelines

The Medical Factors

No Standard of Care

The lack of a clear standard of care to prevent hereditary breast cancer is one significant medical difference between BRCA and other hereditary cancers that may explain why the previvor identity has stayed squarely within the BRCA community [3]. Most hereditary cancers have established prophylactic treatments that are routinely recommended by health care professionals. For example, the standard of care for individuals with gene alterations related to FAP is a prophylactic colectomy between 18 and 20 years old [4]. While there is a standard of care for BRCA positive women, it pertains only to their ovarian cancer risk: surveillance for ovarian cancer has proven ineffective [5], so the consensus is that oophorectomy should be offered to all women carrying a BRCA mutation between age 35-40 or once childbearing is complete [3].

The current options to minimize breast cancer risk are “watchful waiting” (i.e., early detection strategies such as more frequent screenings), prophylactic medication (namely Tamoxifen), or prophylactic surgery (mastectomy and oophorectomy). Although there is strong evidence for the effectiveness of prophylactic mastectomy in preventing breast cancer in BRCA positive women [6], it is not considered standard of care and moreover discussing it as a treatment option is not even considered the standard of care. For instance, the American Congress of Obstetrics and Gynecologists (ACOG) lists the various options for addressing breast cancer risk, but does not state that any of these options should be followed or should be discussed. Yet, ACOG explicitly names the standard of care for ovarian cancer risk: “risk-reducing salpingo-oophorectomy, which includes removal of the ovaries and fallopian tubes in their entirety, should be offered by age 40 years or after the conclusion of childbearing” [7, p.960]. Similarly, the National Comprehensive Cancer Network (NCCN) guidelines do not list a standard of care or require mention of certain treatment options: “Discussion of risk-reducing mastectomy should be carried out on a case-by-case basis” [3]. But only 16% of all physicians follow the NCCN guidelines for treating BRCA positive women [8], so patients receive a range of information about prophylactic mastectomy and potentially no information at all. Even when prophylactic mastectomy is discussed, patients often do not receive decisive advice on the best risk management strategy for them, so the decision to pursue prophylactic mastectomy is often based on individual preferences rather than an established medical protocol [9, p.760;10, p.657]. Specifically, some women’s decisions are based on fear and anxiety, as well as not wanting to live with the guilt of knowing they could have done something but chose not to [11].

Many women with a BRCA mutation want clear and unambiguous advice from their physicians about what treatment options to pursue [12], but without this and without a standard of care to fall back on, they may be left floundering to make the “right” decision and/or the one that most aligns with their values. The existence of an established standard of care relinquishes the burden of the difficult decision to remove an entire body part. The difficulty of the decision of what treatment options, if any, a BRCA positive woman should take distinguishes it from other hereditary conditions where there are either no preventive options available (e.g., Huntington’s disease) or there is a clear standard of care (e.g., FAP).

Genetic Penetrance and Mortality

Another important medical difference between BRCA and other genetic cancers is that the penetrance for BRCA encompasses a large range. Whereas cancer is virtually guaranteed for individuals with

FAP, this is not the case with BRCA carriers: their lifetime incidence of malignancy is 45-85% for breast cancer and 11-62% for ovarian cancer [2]. A penetrance rate of almost 100% is a strong justification for prophylactic surgery. It is harder, however, to support prophylactic surgery when penetrance is a 40-50 point range and it is difficult to know the exact probability of penetrance for a specific individual. The risks and benefits of someone on the low end of a 40-50 point range are very different from someone at the high end.

One important risk to consider when weighing prophylactic surgery is the average mortality rate from the hereditary cancer if one opts not to have the surgery. Here again there is a stark difference between BRCA and FAP. Without surgical intervention, mortality from gastrointestinal cancer for individuals with FAP is almost 100%, with half of patients dying before age 50 [4]. In contrast, individuals with a BRCA mutation who do not have prophylactic surgery can have a normal, or near normal, life expectancy [13]. Part of the reason for this difference is that there are effective, nonsurgical prophylactic treatment options available to BRCA positive women (e.g., Tamoxifen) while there are none for individuals with the FAP mutation. Furthermore, there are also successful treatments for breast and ovarian cancer that can allow for normal life expectancy for women who are BRCA positive and develop cancer. Even if women with a BRCA mutation choose to have no intervention whatsoever, some of them can still be expected to live into their 70s [13].

The Social Factors

Cultural Prominence of Breast Cancer

Breast cancer is one of the most well-known and well supported disease causes. The pink ribbon is a universally recognized symbol of breast cancer and can be found affixed to myriad consumer products of companies that aim, at least in part, to show that they are “woman friendly” [14]. Fundraising and marketing is not the only area in which breast cancer is pervasive; breast cancer is overrepresented in popular press, which fuels women’s anxiety about the disease [15-17]. Many media portrayals of women with breast cancer show and/or describe pre-menopausal women, which can lead women to believe that risk of breast cancer before menopause is high, even though it is not [18,19]. Indeed, some research shows that the majority of women erroneously believe that breast cancer is the number one killer of women, when in fact it is heart disease [20]. Women’s intense fear of breast cancer is part of the reason that breast cancer continues to be overrepresented in the popular press.

The BRCA previvor identity has emerged as yet another breast cancer threat with women mistakenly believing that they are at risk for hereditary breast cancer. The cultural prominence of breast cancer provided a springboard for BRCA previvors; and BRCA previvors have emerged as another narrative within the breast cancer umbrella. Because other diseases (including other types of hereditary cancer, like FAP) do not carry the same degree of cultural currency of breast cancer, previvors of those diseases have a much more difficult time gaining public interest. Without such public interest, it is harder to raise money for research and to support previvors. On the individual level, BRCA previvors may receive more sympathy and support from friends and family than other previvors because BRCA is a well-known disease that is publicly feared. In short, the ubiquity of breast cancer advocacy and media coverage provided the cultural context and foundation for which the identity of BRCA previvors could develop and gain public attention.

Significance of Breasts

Due to mind/body dualism, we often assume that “organs are simply mechanical entities whose worth is entirely without symbolic or affective meaning” [21, p.1408]. When our organs are functioning properly, we typically do not think much about them and any role they may play in our identity. However, certain organs, such as the hands and face, carry more symbolic weight than others [22]. Breasts are another example of organs with symbolic value: they serve as gender markers (i.e.,

identifiers of one's sex) and are generally seen as a central part of women's sexuality, reproductive capacity, and overall femininity. Not surprisingly then, "[t]he fear of loss of femininity, sexual attraction and loss of sexual pleasure" is a significant concern for many women who are considering and eventually undergo prophylactic mastectomy [23, p.793]. While people with FAP also have anxieties related to sexuality following their prophylactic surgery [24], these typically do not have to do with deeper concerns regarding their sexual identity since the colon is not a symbol of sexuality in the way that are breasts. Breasts are more closely linked to one's gendered, sexual, and reproductive self and so the decision to remove them prophylactically is much more complicated and difficult and their loss is felt more deeply than the loss of other organs that are not as intertwined with our identity.

Yet, breasts are important not just to individual women, but also to our society as a whole as they connote important cultural values such as fertility, nurturing/caregiving, luxury, and political freedom [25]. The cultural significance of breasts is reflected in laws, such as the US Women's Health and Cancer Rights Act of 1998, that mandates that if health insurance companies cover the costs of mastectomy for cancer patients, then they must also cover the costs of breast reconstruction for mastectomy patients. No other quality of life laws for cancer patients exist, even for comparable conditions like iatrogenic infertility [26].

Conclusions

In order to gain a deeper understanding of the experiences of women with a BRCA mutation, it is useful to be familiar with the previvor identity as well as both the medical and social factors that shape it. In particular, recognizing what distinguishes women with a BRCA mutation from others with hereditary cancers – i.e., the lack of a medical standard of care, the genetic penetrance and lower mortality caused by the mutation, the cultural prominence of breast cancer, and the significance of breasts – will enable healthcare providers to best support and treat BRCA previvors. In particular, healthcare providers should be cognizant of the difficult decisions that BRCA previvors face due to the absence of established guidelines for handling breast cancer risk. The fact that there is no set standard may be seen as empowering for patients, at least on the surface. In reality, however, patients are often burdened with the responsibility of making such a challenging decision and may face guilt and regret regardless of the decision they make. Continued scientific studies on how to best address breast cancer risk for women with a BRCA mutation is imperative in order to move toward a clear standard of care. In the meantime, healthcare providers should do their best to counsel women and ensure that they are receiving psychosocial support when making these difficult decisions.

List of References

1. Friedman S. Previvor: past, present, & future. FORCE: Facing Our Risk of Cancer. Empowered, July 22 2008 (updated September 28, 2016).
2. You YN, Lakhani VT, Wells SA, Jr. The role of prophylactic surgery in cancer prevention. World J Surg. 2007;31(3):450-64.
3. Daly MB, Axilbund JE, Buys S, Crawford B, Farrell CD, Friedman S, et al. Genetic/familial high-risk assessment: breast and ovarian. Journal of the National Comprehensive Cancer Network. 2010;8(5):562-94.
4. Fritzell K, Persson C, Bjork J, Hultcrantz R, Wettergren L. Patients' views of surgery and surveillance for familial adenomatous polyposis. Cancer Nursing. 2010;33(2):E17-23.
5. Oei AL, Massuger LF, Bulten J, Ligtenberg MJ, Hoogerbrugge N, de Hullu JA. Surveillance of women at high risk for hereditary ovarian cancer is inefficient. British Journal of Cancer. 2006;94(6):814-9.
6. Bermejo-Perez MJ, Marquez-Calderon S, Llanos-Mendez A. Effectiveness of preventive interventions in BRCA1/2 gene mutation carriers: a systematic review. International Journal of Cancer. 2007;121(2):225-31.

7. American College of O, Gynecologists, Bulletins--Gynecology ACoP, Genetics ACo, Society of Gynecologic O. ACOG Practice Bulletin No. 103: Hereditary breast and ovarian cancer syndrome. Obstetrics and Gynecology. 2009;113(4):957-66.
8. Dhar SU, Cooper HP, Wang T, Parks B, Staggs SA, Hilsenbeck S, et al. Significant differences among physician specialties in management recommendations of BRCA1 mutation carriers. Breast Cancer Res Treat. 2011;129(1):221-7.
9. O'Neill SC, Valdimarsdottir HB, Demarco TA, Peshkin BN, Graves KD, Brown K, et al. BRCA1/2 test results impact risk management attitudes, intentions, and uptake. Breast Cancer Res Treat. 2010;124(3):755-64.
10. Printz C. New data on BRCA mutations and prophylactic surgeries. Cancer. 2011;117(4):657.
11. van Dijk S, van Roosmalen MS, Otten W, Stalmeier PF. Decision making regarding prophylactic mastectomy: stability of preferences and the impact of anticipated feelings of regret. Journal of Clinical Oncology. 2008;26(14):2358-63.
12. Klitzman R, Chung W. The process of deciding about prophylactic surgery for breast and ovarian cancer: Patient questions, uncertainties, and communication. American Journal of Medical Genetics Part A. 2010;152A(1):52-66.
13. Sigal BM, Munoz DF, Kurian AW, Plevritis SK. A simulation model to predict the impact of prophylactic surgery and screening on the life expectancy of BRCA1 and BRCA2 mutation carriers. Cancer Epidemiol Biomarkers Prev. 2012;21(7):1066-77.
14. Ehrenreich B. Slap on a pink ribbon, call it a day. Salon. Dec 2 2009.
15. Yadlon S. Skinny women and good mothers: the rhetoric of risk, control, and culpability in the production of knowledge about breast cancer. Feminist Studies. 1997;23(3):645-677.
16. Blanchard D, Erblich J, Montgomery GH, Bovbjerg DH. Read all about it: the over-representation of breast cancer in popular magazines. Prev Med. 2002;35(4):343-8.
17. Clarke JN, Everest MM. Cancer in the mass print media: fear, uncertainty and the medical model. Social Science & Medicine. 2006;62(10):2591-600.
18. Andsager JL, Hust SJ, Powers A. Patient-blaming and representation of risk factors in breast cancer images. Women Health. 2000;31(2-3):57-79.
19. Haas JS, Kaplan CP, Des Jarlais G, Gildengoin V, Perez-Stable EJ, Kerlikowske K. Perceived risk of breast cancer among women at average and increased risk. Journal of Women's Health. 2005;14(9):845-51.
20. U.S. Department of Health & Human Services. Women's fear of heart disease has almost doubled in three years, but breast cancer remains most feared disease. About the Heart Truth, NIH. Feb 29 2012.
21. Lock M. Human body parts as therapeutic tools: contradictory discourses and transformed subjectivities. Qualitative Health Research. 2002;12(10):1406-18.
22. Swindell JS. Facial allograft transplantation, personal identity and subjectivity. Journal of Medical Ethics. 2007;33(8):449-53.
23. Kenen RH, Shapiro PJ, Hantsoo L, Friedman S, Coyne JC. Women with BRCA1 or BRCA2 mutations renegotiating a post-prophylactic mastectomy identity: self-image and self-disclosure. Journal of Genetic Counseling. 2007;16(6):789-98.
24. Andrews L, Mireskandari S, Jessen J, Thewes B, Solomon M, Macrae F, et al. Impact of familial adenomatous polyposis on young adults: quality of life outcomes. Dis Colon Rectum. 2007;50(9):1306-15.
25. Wiseman S. From the luxurious breast to the virtuous breast: the body politic transformed. Textual Practice. 1997;11(3):477-92.
26. Campo-Engelstein L. Consistency in insurance coverage for iatrogenic conditions resulting from cancer treatment including fertility preservation. Journal of Clinical Oncology. 2010;28(8):1284-6.