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SPECIAL TOPIC

Neurofibromatosis Type 1: Review of Cutaneous and Subcutaneous Tumor Treatment on Quality of Life

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Background: Neurofibromatosis Type 1 (NF1) is the most common type of neurogenetic disorder with a worldwide incidence of between 1 in 2,600 and 1 in 3,000. NF1 has a wide range of manifestations; as a result, NF1 has no "public persona." Beginning at puberty and continuing thereafter patients may grow cutaneous and subcutaneous tumors (neurofibromas) in large numbers, which cause severe problems with appearance, which are similar in severity to those of psoriasis. Appearance concerns due to tumors affect NF1 patients' Quality of Life (QoL). NF1 patients are at increased risk for depression and treatment for depression and significantly enhance QoL. Improving appearance and QoL by decreasing the number of tumors is the NF1 patient's greatest concern. Improving QoL is an end in itself.
Methods: There are no currently available medical or pharmacological treatments for cutaneous and subcutaneous tumors of NF1. Surgery is the only treatment option for cutaneous and subcutaneous tumors. High Quantity (HQ) surgical removal by several methods has been used.
Results: HQ electrosurgical (ES) allows removal of large numbers of tumors, hundreds per surgical session, appears to be highly effective, and most patients are happy with their results. Other surgical techniques, such as scalpellic surgery, are not effective. Inadequate insurance payments, few plastic surgeons performing HQ tumor removal, lack of support from nonsurgeon physicians, few if any NF clinics providing HQ tumor removal, pediatric orientation, focus on basic research, and a dismissive attitude toward NF1 patients are all components in the unavailability of HQ NF tumor removal. Such surgery is not cosmetic but restoration of normal appearance from disfigurement due to a congenital, neoplastic disease.
Conclusions: National legislation is required to mandate insurance companies to provide coverage for HQ tumor removal. Payments by health insurance companies to plastic surgeons must be reasonable and comparable with other procedures. Effort by neurologists and other nonsurgeons is needed to gain support of plastic surgeons. Strong pressure is needed by support groups of NF for providing NF1 HQ surgery at institutions with NF clinics and for proper payments for surgeons and others. *(Plast Reconstr Surg Glob Open 2019;7:e1982; doi: 10.1097/GOX.0000000000001982; Published online 18 January 2019.)*

INCIDENCES

Neurofibromatosis Type 1 (NF1) is an autosomal dominant inherited neurological disorder with a 50% incidence from de novo mutations, an equal incidence between males and females, and a worldwide distribution. NF1 is one of the more common rare disorders[1] and is the most common neurogenetic disorder.[2] There are approximately 107,000 to 123,000 people in the United States with NF1, 1 in 2,600 to 1 in 3,000.[3] There are 82,000 to 94,600 adults with NF1 and 24,700 to 28,500 children with NF1, an adult to child NF1 ratio of 3 1/3 to one.

NF HISTORY

Neurofibromatosis (NF) like cases have been pictorially identified since the 13th century. Friedrich Von Recklinghausen provided evidence of neurofibromas in 1882, and this disease became associated with the eponym von

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Recklinghausen's Disease (VRD). Joseph Carey Merrick (1862–1890) was self-proclaimed as The Elephant Man, and the disorder became known as Elephant Man Disease (EMD). EMD was confused with VRD until the gene for NF1 was discovered in 1990, and Merrick's bones tested negative for the NF1 gene. Subsequently, EMD was shown to be the Proteus syndrome, (incidence 1 in 1,000,000), identified in 1979.[4] The popularity of the story of EMD in films and books has increased interest, awareness, and research in NF1 since 1971 when Ashley Montagu published a book called *The Elephant Man: A Study in Human Dignity.* But this has also led to the stigma and misnomer of NF1 being labeled "Elephant Man disease."[4] Much research has focused on the molecular genetics of NF, which has resulted in identification of types, subtypes, and linkage with other disorders; hence, VRD is now NF1.[5]

NF FEATURES AND COMPOSITION

NF1 has a wide spectrum of types, subtypes, manifestations, and severities. NF1 patients may have a variety of manifestations, which vary greatly and with age and sex.[1,6,7] As a result, NF1 has no clear public "persona."[8] There are currently 3 types of neurofibromatosis: NF1, NF2, and NF3 (Schwannomatosis).[4] Diagnostic criteria are listed in Table 1.[1,9,10] Neurofibromas are composed of Schwann cells, fibroblasts, perineural cells, mast cells, axons, and blood vessels.[6] There are 3 growth patterns: localized (nodular, discrete), diffuse, and plexiform.[11] There are 4 groups of neurofibromas: cutaneous (local or diffuse), subcutaneous, plexiform (nodular or diffuse), and spinal.[12] Skin tumors may vary in size from a millimeter to 10 cm or more.[13]

EFFECTS OF NF ON QUALITY OF LIFE

Skin tumors usually begin to grow at the time of puberty and continue thereafter.[1] The effects of skin tumors all over the body have been shown to have a severe and detrimental psychological effect. The skin effects are similar to those of psoriasis, which has significant disability.[14] Psychosocial effects are greater for NF1 patients than life-threatening physical effects.[15] The priority of patients is cosmetic correction and treatment of disfigurement due to the disease.[16] Quality of Life (QoL) studies have been conducted in the last decade only.[15] Studies indicate the importance of improving appearance on QoL in NF1.[14] Voices of patients with NF1 are different and may have an effect on QoL.[14] Other QoL issues include sleep disorders, social isolation, decreased employment opportunities, and the effects of accelerated appearance of aging on NF1 patients.[14] A patient's self-image is closely affected by their perceptions of their doctors', families', friends', and society's view of their condition.[14] Support groups may help patients to cope.[14] No specific research has been done on chronic pain on QoL in NF1 patients, although pain is recognized to be a major determinant of QoL in other populations.[14] There is a generalized decrease in QoL in NF1 patients versus controls.[14]

The "Psychosocial Burden,"[15] the "Specter of Elephant Man Disease" (EMD), effects of confusion of NF1 with Proteus Syndrome,[4] the concept of "Stigma," (1963) and anxiety provoking "What if?" are addressed.[15] For example, the uncertainty of and the progressive nature of the disorder lead to anxiety of being covered in tumors, developing life-threatening complications, being blind, or being unemployed due to the condition. The chief sources of stigma are family, school, workmates, and negative interactions with medical personnel. Physicians have confused EMD with NF1 in informing patients about their diagnosis,[15] and many doctors today mistakenly identify NF1 as EMD (1986, 2011).[15]

The progressive nature of NF1 and lack of treatment options mean that for many people with this condition, QoL may be the most valid outcome measure to assess success of a treatment because QoL is an end in itself.[14]

Table 1. Diagnostic Criteria: NF1,[1] NF2,[9] and NF3[10]

The NIH criteria for a diagnosis of NF1 are 2 or more of the following:
1: Six or more café-au-lait macules over 5 mm in greatest diameter in prepubertal individuals and over 15 mm in greatest diameter in postpubertal individuals.
2: Two or more neurofibromas of any type or one plexiform neurofibroma.
3: Freckling in the axillary or inguinal regions.
4: Optic nerve glioma or optic chiasma glioma
5: Two or more Lisch nodules in iris
6: A distinctive osseous lesion such as sphenoid dysplasia or thinning of the long bone cortex with or without pseudoarthrosis
7: A first-degree relative with NF1 by the above criteria, developed by a NIH Consensus Conference in 1987.[1]

Diagnosis of Neurofibromatosis (NF) Type 2 (NF2)[9]
Confirmed (definite) diagnosis of NF2:
1. Bilateral vestibular schwannomas (may also be known as acoustic neuroma) (Probable diagnosis of NF2:
1. Family history of NF2 and
2. Unilateral vestibular schwannomas or any 2 of the following tumor types: meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacity, juvenile cortical cataract

A person should be evaluated for NF2 if the conditions below are met:
- Unilateral vestibular schwannoma plus at least 2 of any of the following: meningioma, glioma, schwannoma, juvenile posterior subcapsular lenticular opacities/juvenile cortical cataract
- Two or more meningiomas plus unilateral vestibular schwannoma or any 2 of the following: glioma, schwannoma, juvenile posterior subcapsular lenticular opacities/juvenile cortical cataract

Diagnosis of Schwannomatosis (NF3)[10]
A diagnose for schwannomatosis may be established if a person meets one of the conditions identified below.
Confirmed (definite) diagnosis of schwannomatosis:
- Two or more nonintradermal schwannomas (at least 1 with histologic confirmation)
- No evidence of a vestibular tumor on high-quality MRI scan
- No know constitutional NF2 mutation
- One pathologically confirmed nonvestibular schwannoma plus a first-degree relative who meets the above criteria

Presumed (probable) diagnosis of schwannomatosis:
- Two or more nonintradermal schwannomas (at least one with histologic confirmation)
- No evidence of vestibular tumor on high-quality MRI scan
- No known constitutional NF2 mutation
- No symptoms of eighth nerve dysfunction
- Radiographic evidence (image scans) of a nonvestibular schwannoma plus a first degree relative meeting the criteria for definite schwannomatosis

Only a doctor with expertise in schwannomatosis can provide an accurate diagnosis.

MRI, magnetic resonance imaging; NIH, National Institute of Health.

There is a history of dismissal of NF1 patients by the medical profession.[15] There is a lack of family physicians who know about NF1 and scarcity of NF1 clinics specialized for adults.[14]

Few clinicians or researchers have knowledge of the daily life of NF1 patients.[15]

Clinicians believe most persons with NF1 are not and will not be severely affected by their disorder.[15] In 1993, it was found most NF1 patients judge their condition to be more severe than their physicians appreciated.[15] Clinical assessments do not factor in the psychological impact of symptoms such as even quite small NF's, though these affect patient decisions throughout life.[15] NF1 patients' negative self-images typically have resulted from societal insults around their condition (1986).[15] A comprehensive exploration of psychosocial issues in NF1 was published in 1999.[15]

Adult NF1 patients' primary concern is that their tumors, whether visible or not, cause them to be rejected by others. Well-documented cosmetic and social "specifications" are a basis for stigma.[16] Visible stigma cause immediate concern. Nonvisible stigma cause concern when revealed, which can be anxiety provoking. The impact of skin appearance in NF1 has been found to be similar in magnitude to psoriasis.[14] Psoriasis treatment is covered by insurance because it is not a cosmetic issue (improvement in normal appearance) but a medical issue (restoration of a normal appearance from disfigurement) and NF1 is exactly the same. These 2 conditions and others, such as acne vulgaris or vitiligo require medical therapy or reconstructive surgery, not just plastic surgery for cosmesis. It is noted that "bullying" is now a social topic. Patients are bothered by their disfigurement whether tumors are visible or not. "If I have surgery once or twice every few years, people don't shun me for that." Despite large internal tumors, which were fatal in one case, the patient's concern was on disfigurement.[15]

A high rate of appearance concerns among women has been found. Many of these concerns were related to NF and to psychosocial factors such as feeling self-conscious with others. Living with NF may negatively impact women's self-perceptions and underscore the importance of addressing this impact. Research has almost exclusively focused on the medical needs of adult patients with NF, with much less attention on the psychosocial and sexual impact of living with NF. Treatment programs and multidisciplinary clinics that address patients' psychosexual and social needs, including appearance-related concerns, are lacking. Appearance concerns are highly prevalent among women with NF1 and NF2 and may be related to distressing outcomes such as loneliness and reduced self-esteem. At work, appearance, medical problems, and absences for doctor visits or medical treatments are sources of stigma, as are intimacy and marriage.

Protection through government action can improve the QoL via access to health insurance and equal opportunity for employment.[14] The U.S. Genetic Information Non-Discrimination Act of 2009 prohibits use of genetic information for discrimination regarding employment or access to health insurance,[14] though health insurance access alone is insufficient due to the many restrictions and exclusions by health insurance providers.

There has been no investigation of the magnitude of effect of living with this genetically transmissible disease on QoL. No specific research has quantified the effects on QoL of employment, economic difficulties, or the stress of being uninsurable. Such studies could justify governmental intervention.[14]

Cosmetic surgery, speech therapy, education, and behavioral modifications in childhood have the potential to make a lifelong impact on QoL.[14]

MEDICAL TREATMENTS

There are no currently available effective and confirmed medical or pharmacological treatments for cutaneous and subcutaneous tumors of NF1.

PSYCHOLOGICAL TREATMENT—DEPRESSION

Individuals with NF1 are at increased risk for depression and it has been suggested that this population should be routinely screened for depression. Depression was found to be strongly associated with QoL. It is likely that effectively treating depression may significantly enhance QoL for individuals with NF1.[18] Clinicians are urged to assess for psychological, sexual, and social difficulties among patients with NF.[17]

Successful coping involves family support, clear family communications, attendance at NF support groups, and a philosophy that disavows physical differences.[15]

SURGICAL TREATMENTS

Surgery is the only treatment option available for cutaneous and subcutaneous tumors,[6] and in most reports, patients seem to be happy with their surgical results.[3,13,19–25] High Quantity (HQ) surgical removal is an NF1 patient's best option for improving QoL.

Opinions about treatment and removal of cutaneous and subcutaneous NF's differ. Patients are often told NF's are not to be removed, or remove only 5–7 at a time, or that they should learn to tolerate their disorder. Although removing only a few tumors at a time or the most bothersome is beneficial, this does not result in a major change in the patient's condition, outlook or QoL, which removing greater numbers of tumors can have. The traditional approach of removing only larger symptomatic lesions versus early periodic removal of cutaneous lesions to avoid disfigurement and psychosocial stigma is discussed.[25] HQ tumor removal by several methods has been used, but scalpellic tumor removal is impractical.[3] Those few plastic surgeons who are performing HQ removal using several nonscalpellic methods favor this HQ approach (see below.). Most or all patients in several studies of HQ removal have reported positive surgical and QoL results. If these cutaneous and subcutaneous tumors are not removed when they are still small, in some patients their sizes and numbers may increase so much that surgery is no longer an option.

Removal of hundreds of tumors during an operative session has been accomplished with electrosurgery using monopolar Electro-Surgical Units and other methods.[3,19,22,25,26] Use of an Electro-Surgical Units allows performing cutaneous, subcutaneous, and other tumor removals at deeper levels during the same session.[3] Up to 1,000 small tumors or more can be removed during a single session.[25] One operative report indicates removal of 2,700 tumors in 1 session (personal communication). A recent publication about HQ electrosurgery[25] confirms earlier results.[3] Controlled studies have shown no difference in scarring or outcomes between use of an electrosurgical device and a scalpel.[27–29] Electrosurgical techniques are discussed in the following references.[30–38]

Ten photographs from 2008, 2012, and 2018 show a patient (Figs. 1–10) who has had 13 NF1 high-quantity tumor removal operations all over her body since 2008. From 150 to 700 tumors were removed per operation, an average of 340 with an approximate total of 4,100 removed. There has been little or no progression in tumor sizes or numbers in 10 years using a periodic maintenance strategy. Multiple additional patient examples of preoperative and postoperative photographs of HQ NF removal can be viewed online.[39,40]

Surgical management has been shown to be relatively inexpensive from the viewpoint of the healthcare system.[6]

BARRIERS TO HQ TUMOR REMOVAL

Although the recent medical literature documents the advantages of HQ removal, some health insurance companies seem to ignore these articles and rely on outdated literature.[41] Insurance companies discount the value of such surgery and even exclude any discussion of HQ tumor removal; the reasons for these omissions are vague and not consistent with modern techniques and results.[42–44]

Treatment of cutaneous and subcutaneous tumors by electrosurgical removal of neurofibromas in high quantities under general anesthesia appears to be highly effective in improving the psychosocial and QoL issues of NF1 patients. Why is this procedure and similar procedures performed by so few surgeons at so few institutions around the country and not at those with NF clinics? The possible inhibitors include health insurance, payment valuations, procedure codes, being a rare disorder, institutions, NF clinics, pediatric orientation, neurologists, research orientation, anesthesiologists, plastic surgeons, and inadequate pressure by NF1 support groups and patients. Reasons why HQ surgery is not available are listed in Table 2.

Some NF1 patients may lack health insurance, while those who have had the HQ procedure frequently have insurance through employment. Some cases are funded by

Fig. 1. Photograph from 2008 showing the left face before any surgery.

Fig. 3. Photograph from 2012 showing the front after initial surgery and 2 yearly maintenance surgeries.

Fig. 2. Photograph from 2008 showing the right face before any surgery.

Fig. 4. Photograph from 2012 showing the right face after initial surgery and 2 yearly maintenance surgeries.

Fig. 5. Photographs from 2018 showing the front 1 month postoperatively and 8 prior maintenance surgeries.

Fig. 6. Photographs from 2018 showing the left profile 1 month postoperatively and 8 prior maintenance surgeries.

Medicaid. Some surgeons accept insurance and some do not. Insurance companies may pay something but some may reject coverage, classifying the procedure as "cosmetic" or categorizing the tumors as "skin tags" or declaring them not to be "symptomatic." The American Society of Aesthetic Surgeons stated this surgery is clearly "medically necessary" but this organization is focused only on "medically unnecessary" surgery and therefore it has no interest in this procedure (personal communication). The American Society of Plastic Surgeons (ASPS) has stated this surgery is "not cosmetic surgery" (personal communication). The annual statistical report of the ASPS lists tumor removal as "reconstructive surgery," and for 2015 listed 4.5 million operations performed, by far its largest in this category.[45] While cosmesis is a critical reason for this surgery for NF1 patients, the disorder is both neoplastic and symptomatic-physically and psychologically. In addition to symptom relief "restoration of normal appearance" is a key reason for this surgery (as opposed to typical cosmetic surgery's "improving normal appearance"). For some insurance companies to refuse coverage as "cosmetic" for a congenital neoplastic disorder is unacceptable.

Aetna, for example, categorizes symptoms related to "appearance," with few exceptions, as "cosmetic" and

Fig. 7. Photograph from 2018 showing the right profile 1 month postoperatively and after 8 prior maintenance surgeries.

Fig. 9. Photograph from 2018 showing the right profile 8 months postoperatively and after 8 prior maintenance surgeries.

Fig. 8. Photograph from 2018 showing the left profile 8 months postoperatively and after 8 prior maintenance surgeries.

Fig. 10. Photograph from 2018 showing the front 8 months postoperatively and after 8 prior maintenance surgeries.

Table 2. Reasons Stated for Resistance to Surgery Being Available for NF1 Patients

1. "Unaware of the procedure."
2. "Electrosurgery is a major shift from traditional surgery."
3. "This is not a standard procedure."
4. "This procedure will cause scarring."
5. "Plastic surgeons do not like a procedure which causes scars."
6. "Tumors can recur."
7. "There is nothing I can do for you. You are too far gone."
8. "NF1 is a rare disease. Physicians are reluctant to engage in the unknown—scary."
9. "Mindsets, attitudes."
10. "Believe this procedure will always result in loss of function and nerve damage."
11. "Concern about explosion when using a monopolar device requiring grounding of the patient."
12. "Issue of fumes and risk of transmission of viruses."
13. "Doubt NFs are painful."
14. "Dismissive attitude towards NF patients"
15. "Insurance does not pay enough."
16. "Classify as cosmetic surgery."
17. 'Reimbursement is inadequate."
18. "Not worth my time."
19. "There is nothing you can do for this."
20. "A change of heart by the plastic surgeons would be needed for them to perform this surgery."
21. "An unfortunate disorder."
22. "It is all about money."
23. "Many plastic surgeons appear to be unaware of the procedure for NF1 patients."
24. "Remove a few under local anesthesia. Reimbursement is minimal for "add-ons," so there is no incentive to do more."
25. "We will see what happens first, whether you can no longer tolerate this or whether I get bored."
26. "Some think if tumors are removed they will just keep coming back."
27. "Why did the NF clinic refer you to me? I have no interest in this."
28. "Plastic surgeons are interested in complex operative procedures, not simple repetitive procedures."
29. "A Society cannot force plastic surgeons to have an interest in a specific disorder or procedure."
30. "NF is one of many disorders for which plastic surgeons have no interest in performing surgery, and there is nothing the society can do about it."
31. "There are only 5,000 plastic surgeons in the U.S. and they are interested in complex procedures, which this is not."
32. "This surgery would not be challenging and would be mundane and repetitive to most plastic surgeons."
33. "The ASPS has nothing to offer in this regard."
34. An institution with an NF clinic: "Does not perform this surgery because there is not enough payment from insurance companies."
35. "I cannot make enough money doing this."
36. "Opportunity cost, compared to doing other types of surgery."
37. "Not glamorous surgery."
38. "The attitude is, why do anything when new NF's will appear?"
39. "There is plastic surgeon resistance to accept this form of treatment."
40. "Many reasons, logistical and psychological."
41. "Reimbursement is poor for ED or any variant."
42. "Code confusion is part of the problem."
43. "Plastic surgeons lack interest in this area."
44. "A few conditions are neglected by most plastic surgeons, e.g., epidermolysis bullosa and vascular anomalies, "impossible" problems that cannot be taken care of in a single surgery. In fact, there may be a lifetime of surgeries required."
45. "There is a disconnection between plastic surgeons and NF clinics run by neurologists and geneticists."
46. "Poor reimbursements do not justify anesthesia time or purchase of special equipment. The problems are similar in large academic institutions."
47. "Plastic surgeons are overwhelmed and turned off by the extent and number of tumors."
48. "There are problems getting paid by insurance companies."
49. 'NF physicians are more interested in research than treatments."
50. "Reimbursements are typically poor for ED or any variant thereof for the removal of hundreds of tumors, regardless of codes used."
51. "Most surgeons will apply the simpler code and only get fractions of reimbursement after the first tumor is treated."

excluded from coverage. "Psychological" is excluded as a "symptom." CPT Codes for "destruction," 17110, 17111; new Category III CPT Codes 0419T and 0420T, are also excluded from coverage as "cosmetic."[46] United HealthCare excludes all Category III Codes as "experimental."[47] Aetna covers cleft lip, cleft palate, otoplasty, and syndactyly, surgeries that have components of "appearance" and are congenital.[46] Since Aetna allows surgery for "appearance," which is both "congenital" and "cosmetic," why should HQ removal for NF1 patients be excluded? The term "appearance" is nonspecific.

Pediatric neurologists and basic research specialists are typically opposed to tumor removal and HQ removal. Many plastic surgeons are either unaware of the technique of HQ ES removal or opposed for various reasons.

The NF clinics are usually pediatric NF clinics and adults are seen there as well. Almost no major tertiary care institutions with NF clinics (50) are performing HQ removal of NF tumors for NF1 patients, even though other types of surgery for NF1 patients are likely performed.

Tertiary care institutions may decide payments to them are inadequate. Even though these patients are adults, the focus in NF clinics appears to be upon pediatric patients and basic research, both essential but should not exclude this treatment. Adults should be seen in adult neurology clinics or neurology/plastic surgery clinics. Physician awareness of adult NF1 patients would then be increased.

Cosmetic surgery is allowed as a medical deduction by the Internal Revenue Service in certain situations: "You can in-

clude in medical expenses the amount you pay for cosmetic surgery if it is necessary to improve a deformity arising from, or directly related to, a congenital abnormality, a personal injury resulting from an accident or trauma, or a disfiguring disease."[48] NF1 tumors are both congenital and disfiguring.

ACTIONS NEEDED FOR NF1 PATIENTS

Federal legislation, the Women's Health and Cancer Rights Act[49] of 1998, which amended the Employee Retirement Income Security Act of 1974, provided mandatory coverage for breast reconstruction for patients that are insured through self-funded group health insurance plans that are governed by Employee Retirement Income Security Act (group health plans, health insurance companies, and Health Maintenance Organizations as long as the plan covers medical and surgical costs for mastectomy. If individual and group health insurance plans cover mastectomies they must cover reconstructive and other postmastectomy benefits. The law does not apply to Medicare and Medicaid but Medicare covers breast reconstruction if the mastectomy were due to breast cancer. Coverage under Medicaid varies by state.

The NF1 problem should be approached in the same manner as the 1998 Women's Health and Cancer Rights Act law. NF support groups should lobby to do the following: (1) NF physicians need to discuss and refer patients for HQ surgery and advocate and arrange for a local surgeon to perform such surgery. (2) NF clinics' attending physicians should work in the adults' clinic and with pediatric patients. (3) Educate plastic and reconstructive surgeons to be aware of the newer literature on this subject. (4) Lobby for payments to be comparable to other reconstructive procedures.

It is past time to stop telling patients to learn to put up with their condition and to discard the approach of removing specific lesions only, and stating the long-term benefit of removing a large number of neurofibromas is untested. A variety of surgical methods with a high degree of patient satisfaction has moved 2 decades beyond that approach.

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