

Summary of Home Nursing for Lens Dislocation Among Patients with Marfan Syndrome

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Abstract: Marfan syndrome is a rare genetic disease, and the condition of most patients deteriorates with age. The lesions are mainly cardiovascular, skeletal, and ocular lesions. Its clinical manifestations are characterized by congenital heart disease, ectopic lens, slender body, slender limbs, spider fingers (toes), and general muscle dysplasia. In the early stage of this disease, when the symptoms are not yet clearly manifested, some patients came to the hospital for treatment due to decreased vision. According to the particularity of crystal dislocation patients with Marfan syndrome, this article provides home nursing education in terms of physiological and psychological aspects, and builds an effective home nursing model to promote the physical and mental recovery of patients.

Keywords: Marfan syndrome; Lens dislocation; Psychological care; Home care

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1. Introduction

Marfan syndrome is a congenital and hereditary connective tissue disease with an incidence of about 0.04% to 0.1%, is more common in males than females, and is mostly due to autosomal dominant inheritance. Patients with this disease often has family history ^[1], mainly due to congenital dystrophy, mesodermal hypoplasia, and cellulose gene defect. Cellulose is the main component of fibrous filaments or elastic fibers, and is widely distributed in the aorta, zonular ligament, and periosteum of the human body. Hyperextension of connective tissue caused by fibrinogen abnormalities can lead to dilation of the aorta and lens displacement; in skeletal defects, it acts indirectly through the periosteum, which covers the surface of the periosteum and provides counterforce during normal growth. When the elasticity of the periosteum increases, bone hypergrowth will occur ^[2]. It mainly causes cardiovascular, bone and eye lesions, and its clinical manifestations include congenital heart disease, ectopic lens, slender body, slender limbs, spider fingers (toes), and general muscle dysplasia ^[3]. The condition of most patients deteriorates with age. The main morbidity and early mortality are related to cardiovascular disease ^[4], the common ones are aortic dilatation and mitral valve prolapse ^[5]. The common ocular symptoms are ectopia lens, retinal detachment, combined with glaucoma, nystagmus, strabismus, weakness, and many more. Ectopia lens is caused by the laxity or rupture of the zonule, especially the upward and temporal displacement ^[6]. High myopia is common, and some patients experience changes in the angle of the eye chamber. This article mainly discusses ophthalmic diseases and related nursing problems Marfan syndrome patients. By reviewing domestic and foreign literature and summarizing the experience and understanding of the nursing process, the nursing experience of patients with Marfan syndrome diagnosed and treated in the ophthalmology department of our hospital is summarized.

2. Non-surgical treatment home care

Some patients were young, and their family members refused the operation considering the poor cooperation of the patients and the risk of general anesthesia. Besides, the range of lens dislocation was small, so refractive correction and glasses could be given.

2.1. Refractive correction

The doctor's advice should be followed on the wearing of glasses after vision correction is carried through mydriasis with cyclopentolate and eye tests. The patient's family members should strictly supervise the patient's wearing of glasses, avoid various bad eye habits caused by poor vision, and reduce fatigue. At the same time, corrective glasses also an important tool for treating patients with amblyopia. Amblyopia can be effectively treated if the doctor's advice is followed, and the corrective glasses are worn regularly. Some patients are young and need family guidance and supervision. Their daily eye health should be monitored, and they should read books, watch TV, and use other electronic products for not more than half an hour at a time, and should avoid staying in a dark room for a long time. Patients should perform eye massage regularly for 3-5 minutes each time. If the patient experiences eye pain, sharp drop in vision, black vision, and double vision, medical attention should be sought in time. Patients should go for regular check-ups. However, for patients with Marfan syndrome, surgery is the most effective way to treat ectopic lenses in the eye lesions. Therefore, it is necessary to emphasize the importance of surgery for binocular lens ectopia to the family members of the patients, and surgery should be performed as soon as possible.

2.2. Avoid strenuous exercise

Patients with incomplete dislocation of the lens of both eyes should avoid strenuous and extreme sports, such as bungee jumping, diving, football, basketball, lifting heavy objects, defecation, and more. Besides, the patients should avoid emotional agitation and overworking, and mainly eat small and light meals. Collision of the head against other objects should be strictly avoided to prevent eye trauma.

3. Surgical treatment home care

Lens implantation + anterior vitrectomy, surgical treatment of patients is a day surgery mode, that is, the diagnosis and treatment process of patients admitted for surgery and discharged in one working day.

3.1. Condition observation

On the day of the surgery, the eye dressing should be bandaged, and the bandage should not be opened. After two hours of observation, the intraocular pressure should be monitored, whether there is abnormal secretion in the eye, the degree of conjunctival congestion, vital signs, and blood pressure. Postoperative superficial anesthesia and local anesthesia intensified patients should be observed for 1-2 hours. During the observation period, they can eat normally (do not eat spicy food). Patients under general anesthesia need to be observed for 3 hours. Half an hour after oxygen inhalation, they can drink a small amount of water. After 1 hour, they can eat light and digestible food, and the patient needs to be assisted to get out of bed. They can leave the hospital after 3 hours after being evaluated by an anesthesiologist [7]. 24 hours after the operation, observe whether there is any abnormality in the operated eye. Mild grinding, itching, tearing and other symptoms in the eyes after surgery are normal; whereas symptoms such as exudate, blood oozing, edema, pain, and so on, or ocular pain, combined with headache, nausea or vomiting, and so on requires immediate medical attention. On the first day after the operation, the medical staff opens the surgical eye dressing, and then administers eye drops to the surgical eye.

3.2. Medication care

3.2.1. Perioperative eye drops

(1) Antibiotic eye drops, such as levofloxacin eye drops, tobramycin eye drops, gatifloxacin ophthalmic gel; (2) non-steroidal antibiotic eye drops inflammatory drugs, such as pranoprofen eye drops, bromfenac sodium eye drops, diclofenac sodium eye drops; (3) steroid anti-inflammatory drugs, such as prednisolone eye drops, tobramycin and dexamethasone eye drops. 4-6 times a day, with an interval of 5 minutes between medicines. The general time for drug withdrawal is 1-2 weeks after surgery. The specific situation depends on the recovery of the patient after one week of follow-up visit.

3.2.2. Eye pointing method

Trim nails and wash hands before pointing, gently pull the lower eyelid of the patient, drop 1-2 drops from the mouth of the bottle 1-2 cm from the eyelid, gently close the eyes and rest for 3-5 minutes (do not pull on the eyelid) eyelid, postoperative surgical incision on top).

3.3. Life nursing

Protect the eye of the eye from collision, do not squeeze or rub the eye with your hands, and prevent water from entering the eye within 1 month after the operation. Pay attention to rest, ensure adequate sleep; watch TV, use mobile phones, and read books while avoiding eye fatigue; avoid densely populated, dusty, crowded, smoke-stimulated public places, avoid strong light stimulation, and wear sunglasses when going out. Appropriate activities are mainly walking; avoid jumping, running, swimming, and other strenuous activities. Swimming and hot springs are prohibited for three months. Coughing, sneezing, bending over and lifting heavy objects should also be avoided. Moreover, patients should maintain a balanced nutritional intake, eat more vegetables and fruits, crude fiber foods, eat less spicy food, and stop smoking and consuming alcohol to prevent constipation^[8]. The doctor's advice should be followed, and patients should go for check-ups regularly. In addition, patients are advised to visit an ophthalmology clinic for treatment if they experience any eye discomfort, such as eye redness, swelling and pain, and vision loss.

4. Psychological care

4.1. Psychological care of patients

Patients are often stressed, sensitive, and fragile, they might also experience anxiety, depression, and other psychological issues, which has a detrimental impact on their daily lives and education. Therefore, the following measures should be taken: (1) family members should ensure that the patients room is warm, comfortable, and quiet, so that the patient can live in a relatively peaceful state. (2) inform the patient about the relevant knowledge of the disease and the impact of negative emotions on his condition, and encourage the patient to express his inner feelings and vent his negative emotions; (3) family members should accept the patient's negative emotions, actively enlighten them, and pay close attention to the patient's psychological conditions; (4) patients may be at risk of being bullied or marginalized at school or at work due to their body size and facial appearance. Close attention should be paid to the psychosocial risks associated with Marfan syndrome, and the patient needs to be managed appropriately; (5) advise patients to pick up an art-related hobby to allow them to vent their negative emotions, such as piano, violin, and singing.

4.2. Psychological care for family members

Marfan syndrome is a rare disease, and it causes a heavy financial burden and psychological pressure on the family members, which then lead to symptoms such as anxiety, depression, paranoia, and stress among the family members^[9]. Parents of patients need time and space to deal with negative emotions and adapt

to the situation. Therefore, while caring about the psychological conditions of patients, we must also pay attention to the psychological changes of the family members of patients^[10]. Effective guidance should be provided to alleviate the psychological pressure of parents of patients with Marfan syndrome: (1) comprehensively assess the needs and understanding of family members towards Marfan syndrome, and provide one-on-one intensive education on the issues that family members are worried about to alleviate their anxiety; (2) understand the economic conditions of the patient's family, assist in solving the financial difficulties encountered, seek economic support and help from the society and the media, and help them discover relevant social support organizations; (3) family members should be encouraged to join a Marfan syndrome group and seek support from their peers; (4) inform the family members of the patient of possible complications and prognosis during treatment, so that the family members have a positive psychological response to the treatment and are able to stabilize the patient's emotions; (5) actively provide psychological counseling to encourage them to express their psychological feelings, and accept the patient's condition, so that patients can feel safe.

5. Genetic counseling

Marfan syndrome is a progressive disease involving multiple systems. Inform family members of the importance of regular monitoring and follow-up. Consultation should be provided for family members on a regular basis, and they should be educated about Marfan syndrome, and closely observe adverse reactions. Besides, genetic counseling should be provided to understand the needs and concerns of patients' families upon the diagnosis Marfan syndrome disease, which involves carrying out a consanguinity analysis and diagnostic tests to identify family members at risk, and explaining the development process and inheritance mode of Marfan syndrome to patients and their families. If the patient or family members want to have children, genetic counseling, prenatal diagnosis, and decision-making discussions should be carried out^[11].

Marfan syndrome is an autosomal dominant hereditary systemic connective tissue disease^[2], but the pathogenesis is still unclear. The difference in phenotypes indicates that the onset may be related to environmental factors other than genetics. But what kind of environmental factors cause patients to express the huge difference of the phenotype and its molecular basis have become the key points of further research. At present, the prevention and treatment of Marfan syndrome and disease detection in China are mainly based on the evaluation of clinical phenotypes. Early diagnosis based on molecular biology and high-throughput sequencing have become the mainstream methods for the screening of genetic diseases. Therefore, genetic screening should be carried out based on phenotypic assessment for patients and family members who are at risk of developing the disease. Then, biological intervention, gene therapy, tissue engineering can be carried out to repair connective tissue defects and drug treatment of the pathogenesis of Marfan syndrome. The research on the target of action provides a new scientific basis for clinical application of drugs, biological therapy, and surgical therapy^[12], so as to prevent missed diagnosis and the progression of disease, which would be difficult to treat.

6. Discussion

Rhegmatogenous retinal detachment in Marfan syndrome can occur in one or both eyes, and it is more common among young men, with an average earlier age of onset^[13]. Once a retinal hole forms, it usually develops rapidly, leading to multiple holes or huge holes, mostly located in the peripheral part of the retina, often accompanied by vitreous preretinal hyperplasia^[14]. Most of the ophthalmology patients who come to the hospital for treatment have obvious ocular lesions. Therefore, in the process of ophthalmology consultation, in addition to paying attention to ocular symptoms, it is also important to pay attention to the general physical signs, and further guide patients on other characteristic manifestations. When the patient has eye lesions such as ectopia lentis combined with characteristic changes in skeletal muscle and

cardiovascular disease, the patient is advised to undergo further examination. If all aspects meet the relevant symptoms, the patient can be recommended for further genetic testing to confirm the diagnosis of Marfan syndrome. The outpatient cases of the ophthalmology department of our hospital were collected, and patients with abnormal bone changes (including abnormal metacarpal index and flat chest), eye changes (high myopia and lens subluxation). The diagnostic criteria for Marfan syndrome are changes in the cardiovascular system and at least two of the four abnormalities in the family history ^[1]. About 80% of patients diagnosed with Marfan syndrome have clinical manifestations of ectopia lentis. Therefore, when other clinical symptoms are not obvious in the early stage, ocular lesions combined with physical characteristic lesions of Marfan syndrome will aid in its diagnosis.

Home care refers to nurses going to patients' homes to provide personalized and professional nursing services to patients for better disease prevention, health promotion and health maintenance. Foreign home care has been widely used in rehabilitation care, palliative care, geriatric care, maternal and infant care, wound care, and many more. It was found that foreign home care can reduce national medical and health expenditures and improve patient satisfaction ^[15]. It is necessary to provide corresponding home nursing care to prevent frequent hospital visits, which is time consuming and costly. There is no invasive treatment and operation in the ophthalmic care of the disease, and it is completely possible to guide the family members of the patient to monitor and care for the patient at home. At the same time, keeping in touch can provide targeted answers and care for patients' eye problems in a timely manner.

Marfan syndrome is a rare disease. Therefore, while treating the disease, a bio-psychological-social medicine model to address the psychological problems of patients and their families, and provide corresponding psychological care and intervention measures to aid the treatment and rehabilitation of patients. This disease is mainly treated by surgery. The disease treatment, psychological counseling, and intervention for patients and their families should be improved, and establish a good relationship with patients, strive for the cooperation of patients and their families, and perform surgery as early as possible ^[16].

In summary, the introduction of personalized psychological nursing and home nursing in the ophthalmic nursing of patients with Marfan syndrome lens dislocation can reduce the negative emotions of patients and their families, help maintain a good psychological state, and further promote the physical and mental health of patients.

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Disclosure statement

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