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Nursing Implications for the Management of Lymphatic Malformation in Children

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Abstract

Lymphatic malformation (LM) is a benign congenital childhood growth that presents primarily at birth, with the remainder becoming evident by age 2 years. LM can cause devastating complications such as respiratory failure, dyspnea, dysphagia, organ compression, and nerve compression. Surgery is the preferred treatment option, although resection is not always an option due to the anatomic location of the malformation. Other treatments that have been tried with success include sclerotherapy, radiofrequency ablation, propranolol, and sirolimus. Nurses have an important role in caring for the child with LM, as with any complex disease. Nurses provide care at the bedside, education to the family, case coordination, and psychosocial support. LM is a rare disease in infancy necessitating pediatric nurses to support families through prolonged treatments and potential complications.

Keywords

lymphatic malformation, pediatrics, nursing

Introduction

Lymphatic malformation (LM) is a benign congenital lymphatic childhood growth that occurs due to an abnormality between the venous system and the lymphatic system (Han et al., 2011). It consists of cysts of varying sizes, which cause masses (Erikçi et al., 2013; Impellizzeri et al., 2010; Niramis, Watanatittan, & Rattanasuwan, 2010; Perkins et al., 2010). About 60% to 65% of the cases are diagnosed at birth, with 80% to 90% becoming evident by age 2 years (Erikçi et al., 2013; Han et al., 2011; Impellizzeri et al., 2010; Kumar et al., 2012). Occasionally, patients are diagnosed in adulthood (Goswamy, Penney, Bruce, & Rothera, 2013). LM is relatively rare, happening in anywhere from 1 in 1000 to 1 in 16 000 live births (Kumar et al., 2012). It accounts for 6% of benign lesions in childhood (Impellizzeri et al., 2010). Treatment can be challenging, especially for large lesions. There is not one single treatment option for all patients (Goswamy et al., 2013). Surgical excision is the treatment of choice, but it is not always possible due to the malformation's location near nerves and other vital structures (Erikçi et al., 2013; Impellizzeri et al., 2010; Niramis et al., 2010). Other possible treatments include sclerotherapy and radiofrequency ablation, and, recently, propranolol and sirolimus have been used (Erikçi et al., 2013; Goswamy et al., 2013; Impellizzeri et al., 2010; Lackner et al., 2015; Reinglas, Ramphal, & Bromwich, 2011). This article will further explore the treatments available to children with LM and the nursing roles not caring for children with this complex disease.

History and Review of the Literature

Classification and Staging

The International Society for the Study of Vascular Anomalies (ISSVA) has a classification system for all vascular anomalies (see Table 1). Anomalies are classified as either vascular tumors or vascular malformations, based on radiographic findings. Vascular malformations, which are always benign, can be simple, combined where 2 or more types of malformations are found in the same lesion, or associated with other anomalies. Simple malformations include capillary malformations, venous malformations, LM, arteriovenous malformations, and arteriovenous fistulae. Associated anomalies with LM include Klippel-Trenaunay syndrome, CLOVES syndrome, and Proteus syndrome, among others. Vascular tumors, the other category of vascular anomaly, can be benign, locally aggressive, or malignant. These include infantile hemangioma, congenital hemangioma, Kaposi sarcoma, and angiosarcoma among others (ISSVA, 2014).

Three fourths of cases of LM occur in the head and neck region, followed by 20% in the axilla and 2% in the inguinal region (Kumar et al., 2012). LM can be categorized radiographically based on the size of the cysts making up the malformation. LM can be macrocystic (cysts >1 cm),

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Table 1. ISSVA Classification for Vascular Anomalies[a].

Vascular Anomalies						
Vascular Tumors			Vascular Malformations			
Benign	Locally Aggressive	Malignant	Simple	Combined	Of Major Named Vessels	Associated With Other Anomalies
Infantile hemangioma	Kaposiform hemangioendothelioma	Angiosarcoma	Capillary malformations	Capillary venous malformation	Affects lymphatics, veins, arteries	Klippel-Trenaunay syndrome
Congenital hemangioma	Retiform hemangioendothelioma	Epithelioid hemangioendothelioma	Lymphatic malformations	Capillary lymphatic malformation		Parkes-Weber syndrome
Tufted angioma	Papillary intralymphatic angioendothelioma (PILA)	Others	Venous malformations	Lymphatic venous malformation		Servelle-Martorell syndrome
Spindle-cell hemangioma	Kaposi sarcoma		Arteriovenous malformations	Capillary venous-lymphatic malformation		Sturge-Weber syndrome
Epithelioid hemangioma	Others		Arteriovenous fistula	Capillary lymphatic-arteriovenous malformation		CLOVES syndrome
Pyogenic granuloma				Others		Proteus syndrome
Others						Others

[a]Adapted from ISSVA (2014).

microcystic (cysts <1 cm), and of mixed type in which one lesion has cysts of both sizes (Goswamy et al., 2013; Impellizzeri et al., 2010). Histologically, the types of lesions are identical. However, microcystic lesions tend to occur within the mucosa and recur more frequently after treatment compared with macrocystic lesions. The treatment of the different types are different as well, as macrocystic malformations tend to respond to surgery and sclerotherapy, while microcystic malformations do not (Kim et al., 2011). There are 3 classifications of LM: cystic hygroma, lymphangioma simplex, and cavernous lymphangioma. Cystic hygroma, a macrocystic lesion, occurs most often in the head and neck and is the most common subtype in children. It is composed of multiple large cysts filled with serous fluid, which appears as a swelling under the skin. Cystic hygromas are at risk of infection, which can be life threatening due to their location on the head and neck. These are more common in congenital disorders. Lymphangioma simplex is composed of small, thin lymphatic channels and is located in the epidermis. Cavernous lymphangioma, formed of dilated lymphatic channels, frequently invade surrounding tissues. It does not have cystic formation. It more commonly presents in the lip, tongue, cheeks, and floor of mouth (Niramis et al., 2010). Malformations are staged from Stage I to Stage V, based on the relationship to the hyoid bone and cervical laterality (see Table 2). Staging helps determine operative risks and outcome (Perkins et al., 2010).

Table 2. Staging of Cervical Lymphatic Malformations.

Stage	Cervical Laterality	Location to Hyoid Bone
I	Unilateral	Infrahyoid
II	Unilateral	Suprahyoid
III	Unilateral	Infrahyoid and suprahyoid
IV	Bilateral	Infrahyoid
V	Bilateral	Infrahyoid and suprahyoid

Although it is a benign disease, LM can have numerous complications and be life threatening. Malformations present as a painless, asymptomatic mass. They swell as the child grows and with factors such as trauma, infection, and bleeding. As the LM enlarges, there is increased pain, bleeding, and risk for infection. Children with bilateral and microcystic LM have significant lymphocytopenia. The infections cause the malformations to further grow, which complicates the treatment process and leads to further hospitalizations for aggressive antibiotic therapy. Malformations can cause functional compromise and can grow large enough to compress organs. Due to its growth within lymphatic channels, LM is infiltrative and has the potential to grow within nearby structures. Complications of head and neck malformations include respiratory failure, dyspnea, dysphagia, dysphonia, and poor oral hygiene. For patients with LM within the oral cavity, basic oral hygiene causes pain, bleeding, and swelling, which

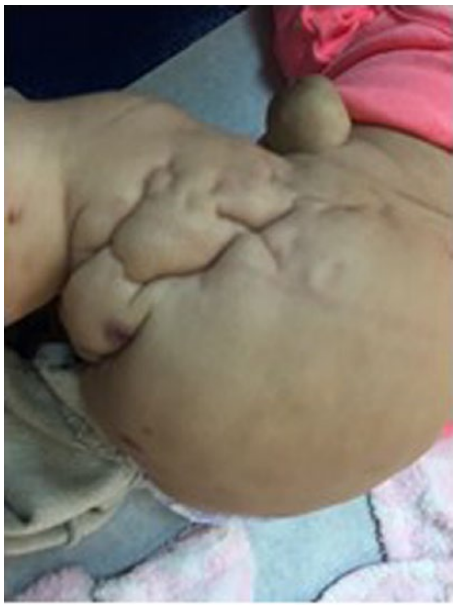

Figure 1. Four-month-old diagnosed at birth with large, left lower extremity LM. The malformation extends into the abdomen causing renal obstruction, which necessitated bilateral nephrostomy tubes and at presentation caused feeding intolerance.

causes the malformation to further enlarge. This puts patients at risk for airway compromise. Providers can educate parents about frequent, gentle brushing with a soft bristled toothbrush. Patients should have their teeth cleaned regularly by a professional, usually more frequently than other children at the discretion of their provider. For patients who have a cervicofacial LM, airway compromise is a frequent concern and is initially treated aggressively with steroids and antibiotics. If these do not control the airway compromise, many patients require a tracheostomy. However, tracheostomies pose a challenge, as they cause chronic airway inflammation, which is a trigger for LM growth. For patients with extensive disease, gastrostomy may be necessary to facilitate feeding. Additionally, all malformations can lead to nerve compression and intracystic hemorrhage (see Figure 1; Han et al., 2011; Impellizzeri et al., 2010; Kim et al., 2011; Niramis et al., 2010; Perkins et al., 2010).

Treatment Approaches

The treatment for LM is challenging. In Stage I and Stage II malformations, the initial treatment is usually curative. In larger malformations, long-term management is required (Perkins et al., 2010). Malformations rarely spontaneously regress, but for small, asymptomatic disease a wait and watch approach is preferable (Han et al., 2011; Kim et al., 2011; Perkins et al., 2010). Additionally, for malformations that have no functional or life-threatening impairments, treatment can be delayed beyond infancy (Perkins et al., 2010).

Surgery is the first line of treatment for macrocystic disease with cysts more than 5 cm (Han et al., 2011). It is less effective for microcystic disease due to the obscuring of normal anatomy (Perkins et al., 2010). A full surgical resection is ideal since there is a high risk of recurrence with partial resection (Impellizzeri et al., 2010; Niramis et al., 2010). Risks associated with surgical resection are due to the malformation's location along the nerves with the potential for injury to vital organs. Additionally, there is a risk for bleeding, wound infection, and scar formation (Impellizzeri et al., 2010; Niramis et al., 2010). Surgery for LM, especially in the cervical area, can substantially affect the cosmetic appearance of children. Recently, some surgeons are moving toward endoscopic surgeries through the neck or chest with better cosmetic outcomes for the treatment of macrocystic cystic hygroma. The standard surgical approach is open, in which a large incision is created for the surgeon to insert instruments and then it is closed with sutures. This has greater risk for morbidity than newer endoscopic surgeries (Han et al., 2011).

Sclerotherapy is used as an alternative and sometimes as an adjunct to surgery for malformations that are nonresectable. Sclerotherapy is when a medication is injected directly into the cyst to try to shrink the malformation. Each agent works slightly differently, but ultimately, all cause local inflammation within the lymphatic channels of the cyst leading to necrosis, while sparing the surrounding structures. This causes the cyst to involute on itself. Sclerotherapy is more beneficial for macrocystic than microcystic lesions (Impellizzeri et al., 2010). Treatment is performed under local anesthesia or general anesthesia depending on the age and cooperativeness of the child and the preference of the provider. First, the provider aspirates contents out of the cyst; then, he or she injects the sclerosing agent into each cyst of the malformation. The patient is observed as an outpatient for up to 24 hours postprocedure. It can be repeated in 2 weeks if the cyst persists and is at least 1 cm in diameter (Kumar et al., 2012; Niramis et al., 2010; Yoo et al., 2009).

There have been multiple sclerosing agents identified in children, with varying degrees of clinical efficacy. OK-432, bleomycin, doxycycline, and sterile ethanol have all been tried with success. The choice of the agent depends on the institution and the provider. Bleomycin, one such agent, is an antineoplastic that irritates the endothelial cells of the cyst wall. Minor side effects include skin erythema, local swelling, induration, and fever, which subside within a few days of injection. The

greatest risk factor of bleomycin is pulmonary toxicity, which is a dose-related side effect, usually occurring only at cumulative doses above 400 mg or at single doses above 30 mg/m^2. Typical doses used for sclerotherapy are lower, ranging from 0.3 to 3 mg/kg (Kumar et al., 2012; Niramis et al., 2010). In one study of 70 children treated with bleomycin sclerotherapy for cystic hygroma, 47.1% had a complete clinical response and 35.8% had greater than 50% reduction of their malformation. The remainder had poor response. In the same study, 3 children died within the first 5 years after injection with bleomycin, of whom 2 were initially poor responders. One died 1 month after the second injection from septicemia and pneumonia, and the second patient needed repeat surgery after injection for a growing LM, who then died from wound infection and septicemia. The third, who was an excellent responder, developed thyroid carcinoma and died from postoperative bleeding after radical neck dissection (Niramis et al., 2010). In another study of 17 children treated with intralesional bleomycin for LM, 35.7% had excellent response and 50% had good response. The remainder had poor response. There were no serious adverse reactions or recurrences during the 18-month follow-up (Erikçi et al., 2013).

OK-432, or picibanil, a mixture of low-virulence Group A *Streptococcus* and penicillin G potassium, a sclerosing agent originally from Japan, has recently gained popularity in the United States. There are 2 theories for its mechanism of action: (*a*) that it induces cytokines, such as *interleukin*-1, *interleukin*-2, and tumor necrosis factors, which recruit inflammatory cells, such as neutrophils, macrophages, lymphocytes, and T-cells to the malformation, and (*b*) that it induces apoptosis of the endothelium of the lymphatic system. It has very few side effects and has shown to be good for both short-term control and long-term control of LM. In a recent study of 55 children with cervical LM, 83.5% showed good short-term response and 76.3% showed good long-term response (Yoo et al., 2009). Further research studies are being conducted in the United States to obtain approval from the Food and Drug Administration for its usage (Perkins et al., 2010).

Doxycycline, a broad-spectrum antibiotic, has been used alone or in combination with ethanol as a sclerosing agent. It creates inflammation and fibrosis within the cyst, similar to other sclerosing agents. For cysts smaller than 3 cc in diameter, it is injected and left for variable amounts of time. For cysts larger than 3 cc in diameter, doxycycline is injected through a pigtail catheter and removed 6 hours later. It is repeated daily for 3 days. Complications include local infection, local inflammation, pain, tooth discoloration in young children, and electrolyte abnormalities (Perkins et al., 2010). In one study of children with macrocystic head and neck LM, 67% of children had complete or near-complete resolution of their lesions after doxycycline sclerotherapy, with a mean follow-up of 1243 days. The remainder, 33%, had no improvement initially or developed recurrence after responding (Jamal et al., 2012).

Sterile ethanol is another sclerotherapy agent that has recently been studied in clinical trials. The mechanism of action is not well understood. It has previously been used for adults and children with arteriovenous malformation and had success in a clinical trial of 8 children with cervical LM who did not previously undergo surgical resection. Of all the patients, 87.5% had a complete response. There were no recurrences after 2 years of follow-up. It has minimal side effects and is inexpensive. Possible side effects include skin and nerve injuries, hemoglobinuria, and possible cardiovascular events (Impellizzeri et al., 2010).

Radiofrequency ablation is a treatment useful for debulking microcystic disease. It is especially useful for symptomatic disease in the tongue and oral cavity, including recurrent bleeding and infection unresponsive to antibiotics (Kim et al., 2011). Radiofrequency ablation leads to reduced symptoms and improved function, in an area where other treatments have the potential to cause significant morbidity. The high-frequency mode destroys deep tissue without affecting the mucosal surface (Kim et al., 2011). The low-frequency mode generates a field of ionized sodium molecules, leading to the removal of target tissue. This technique avoids bleeding and thermal damage that lead to fibrosis and scarring that can be caused by other treatment modalities (Goswamy et al., 2013). In a case series of 5 children with oral and laryngopharyngeal disease, all patients had improved oral intake and respiratory symptoms after undergoing radiofrequency ablation (Goswamy et al., 2013). In a separate study of 26 children with localized, superficial microcystic LM, only one child had complications from the procedure necessitating a stay in the intensive care unit, which resolved quickly. Half of the patients had no symptom complaints at follow-up, and 31% had only minor complaints at follow-up. For the 19% of patients who had continued symptoms disrupting their quality of life, additional radiofrequency ablation was planned (Kim et al., 2011).

Sirolimus, or rapamycin, has been used in small studies of children with various vascular malformations with success. Sirolimus is an inhibitor of the mammalian target of rapamycin, which is involved in cell proliferation and angiogenesis. Mammalian target of rapamycin is overexpressed in LM, which leads to its growth. Sirolimus is frequently used as an immunosuppressant for children receiving renal transplants (Lackner et al., 2015; Reinglas et al., 2011). In one study of 6 patients with various vascular anomalies treated with sirolimus, 3 patients had complete response and 3 had partial response. One patient

who had a partial response was able to have a surgical resection at the completion of treatment. Four were heavily pretreated prior to receiving sirolimus. All the patients were given 0.05 mg/kg orally twice daily and adjusted to achieve serum levels of 5 to 15 μg/L. The only side effect was mild, reversible leukopenia in all the patients (Lackner et al., 2015). In a case report, a 4-month-old with diffuse lymphangiomatosis was treated with sirolimus at a starting dose of 0.4 mg/m^2 up to a maximum of 1.2 mg/m^2. Over a period of 10 months, his malformation decreased in size. Sirolimus levels were maintained between 5 and 10 μg/L. The only adverse effect was mild hypertension, which was controlled with amlodipine (Reinglas et al., 2011).

In a recent study of 6 children with microcystic and macrocystic LM, oral propranolol was studied for effectiveness in decreasing growth size. Propranolol is a nonselective β-blocker used in the treatment of many cardiovascular diseases, and has previously been used in the treatment of infantile hemangioma. Propranolol works by reducing expression of vascular endothelial growth factor, which is high in patients with LM. Children were given 2 mg/kg/day. Patients with gadolinium-enhanced lesions showed a greater response to therapy. Out of the 6 patients, 2 had reduction in the size of their malformation and 2 had symptomatic relief, although their malformation did not shrink. Side effects were minimal, causing only transient lightheadedness and headaches in adolescents (Ozeki et al., 2013). Propranolol should be used with caution in patients with asthma, since it may cause bronchospasms. It should also be used with caution in patients with diabetes, since it can mask the symptoms of hypoglycemia.

Treatment of LM is individualized for each patient, depending on factors such as functional compromise, the stage of the malformation, and the age of the patient. There are no standard treatment protocols for children with LM; however, the American Academy of Otolaryngology-Head and Neck Surgery Foundation published guidelines for treatment planning and prognostication (Perkins et al., 2010). Patients with Stage I, Stage II, or Stage III disease without functional compromise are eligible for observation, single-stage elective surgical excision, or single- or multistage elective sclerotherapy. If patients have functional compromise, they must be treated with surgery or sclerotherapy. All patients with Stage IV and Stage V disease must have long-term management, regardless of the treatment they undergo. Patients without functional impairments may have observation, multistage elective sclerotherapy and surgery, or multistage elective surgery. Patients with functional compromise will have either multistage surgery or multistage sclerotherapy. Additionally, they may have issues with feeding and airway that need to be addressed (Perkins et al., 2010).

Discussion

Physical Needs

LM is a complex disease, which may necessitate long-term treatment beginning in infancy. The pediatric nurse has a role in all aspects of patient care, including bedside care, psychosocial support, education, case coordination, and anticipatory guidance. Patients with extensive disease may be hospitalized frequently. Children with cervical disease have the potential to decompensate due to compression of their airway and need astute monitoring by bedside nurses. Additionally, children may require medical devices such as tracheostomy tubes, gastrostomy tubes, and nephrostomy tubes, depending on the location of the LM. Patients will not only require inpatient monitoring, but they also may require home nursing due to their complex needs. Nurses monitor for acute and long-term side effects of the various therapies patients undergo, some of which require short hospital stays and some of which may require longer hospital stays. Nurses help prepare the pediatric patient for the various treatment therapies that will be utilized, as many of them will be done under anesthesia, and monitor them when they return, including providing adequate pain management.

Children with LM of the head and neck have complex dental issues. Basic oral hygiene can be uncomfortable and may cause bleeding of soft tissues. Patients require frequent dental visits to prevent caries and maintain good oral hygiene. Periodontal inflammation and caries can lead to swelling of the oral cavity, which can compromise the airway. Additionally, children may require orthodontic appliances and jaw surgeries as they get older (Perkins et al., 2010). Nurses can educate families about proper oral hygiene, including the use of a soft bristled toothbrush. Additionally, nurses can ensure that families are following up with dentists at least every 6 months and make appropriate referrals when needed. Children may also have problems with feeding, swallowing, and speech. The nurse helps maintain the nutrition, growth, and development of the child with LM. Nurses involve other members of the team, such as speech therapists and nutritionists to help with these complex issues. From a young age, children with oral LM may have feeding difficulties, either with breast feeding or with oral feeds. They may require nasogastric or gastric tubes. Nurses are responsible for placing and caring for the tubes and educating the family on how to care for them at home.

Psychosocial Care

Parents of children born with LM have a unique set of psychosocial needs. LM is diagnosed in infancy and can cause significant cosmetic deformity, devastating young

parents. Older children and adolescents with physical differences may find school and friendships difficult. Adolescents, especially, may be embarrassed about a large malformation in a visible part of their body. Nurses are able to provide support and refer young patients to outside resources when necessary. Additionally, giving birth to an infant who will require frequent treatments for a chronic illness creates fear and anxiety. As with other chronic illness, parents may miss a lot of time from work creating a financial strain. Nurses can work with case managers to refer eligible patients for medical assistance early after diagnosis. Parents may struggle to find day care centers and preschools that accept their children with special health care needs. Nurses can work with case managers to ensure that families find safe day cares. Parents may have other children, and it can be difficult for them to split time between the hospital and the responsibilities at home. Parents may also need help in explaining their child's condition to their other children. Families need social support from the extended family and the community due to the chronic nature of this disease and treatment. Nurses are positioned to identify families at risk and to make appropriate referrals to psychologists, child life specialists, social workers, and chaplains. Additionally, although therapies are improving for LM, many children continue to have disease with morbidity and mortality. The trajectory can be variable for every child diagnosed with LM, which can be difficult for parents who do not know what to expect at diagnosis in terms of treatment course and eventual outcome. However, nurses can see families through all their treatments and provide a sense of stability and support. LM is a rare disease, and parents may struggle to find other parents going through the same thing. Nurses are in a position to find resources and identify support groups.

Education

Education is vital and is an important part of the nursing role. Families are unlikely to be familiar with LM. Education to families about the disease, the typical course, treatment options, and possible complications is imperative. Families often question the cause of their child's disease. Nurses can explain the pathophysiology of the disease and reinforce that it is not familial and that they did not do anything to cause it.

Children undergoing treatment with specific agents to shrink the malformation will require many medications throughout their treatment. Education should be about the medication, why it is being given, how it works, expected side effects, and discharge planning. Sclerosing agents have the potential to cause localized pain, erythema, swelling, and infection. Although rare, parents of children who receive bleomycin should be warned about the possibility of pulmonary toxicity as a lifelong effect. The nurse should inform them that they should notify all current and future health care providers about the history of bleomycin. Parents of children receiving propranolol should be educated about the possibility of lightheadedness and syncope. They should be instructed to drink enough fluids and return to clinic if symptoms do not subside. Sirolimus causes leukopenia, which increases patients' risk for infection. Parents need education about the signs and symptoms of infection, including fever guidelines, and know when to call or return to clinic. Additionally, patients on sirolimus will require monthly lab draws to monitor their complete blood count and drug levels and for frequent monitoring of triglycerides due to the possibility of hypertriglyceridemia. Sirolimus must be given at the same time every day and cannot be given with grapefruit juice. Additionally, since the majority of patients are babies and toddlers, parents may need advice on how to give medications.

Families require anticipatory guidance about the disease and its chronic nature. Parents need guidance about normal infant milestones and care, in addition to differences due to the malformation. Infants may not meet some milestones on time such as speaking, eating, or walking, depending on where the malformation is located. Additional anticipatory guidance includes when to introduce solid foods, which is individualized for each infant, and the early, frequent need for dental care.

Coordination

Case coordination is required, especially for children with complex disease. Children see a variety of specialists depending on the location of the malformation. Many of these children require multidisciplinary care including obstetrics-gynecology if it is diagnosed prenatally, neonatology, surgery, otolaryngology, oncology, dentistry, nursing, nutrition, speech and swallow, rehabilitation, social work, and child life. Nurses facilitate communication between providers and act as a liaison for families to ensure accurate information is provided. Prior to discharge, the nurse prepares the family for caring for the child at home and the necessary follow-up appointments. Some infants require home health nursing and medical equipment once discharged, which require coordination.

Future Directions

Currently, there is no standard treatment for LM. Further research is needed to find better treatment options when surgery is not curative. Research is needed to find treatments that limit the cosmetic and functional morbidity on patients. Additionally, there is a need to develop standard protocols for patients with LM. Propranolol showed

promising results in one study and can be investigated further for its role in the treatment of LM. Sirolimus can be studied further in clinical trials in the future to assess its impact in the treatment of LM.

Summary

LM is a rare benign malformation in childhood, primarily occurring on the head and neck and mostly affecting infants. Surgery is the primary treatment, although it is not always feasible for malformations that are pressing on nerves and other vital structures. Other treatments include sclerotherapy, propranolol, sirolimus, and radiofrequency ablation. Nurses have a role in caring for the patients at the bedside, providing education to the families, functioning as a care coordinator, and providing psychosocial support. Future directions include combining several treatment modalities at diagnosis, creating standard protocols for patients, and studying the role of sirolimus.

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