



INTERNATIONAL  
LYMPHOEDEMA  
FRAMEWORK

Focus  
Document



INTERNATIONAL LYMPHOEDEMA FRAMEWORK  
**CARE OF CHILDREN WITH  
LYMPHOEDEMA**

[www.lympho.org](http://www.lympho.org)

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# CONTENTS

<b>1. Introduction</b> .....	p.3
<b>2. Childhood lymphoedema: syndromes and presentations</b> <i>Adapted from an original article by Fiona Connell, Consultant Geneticist at South East Thames Regional Genetics Service, Guy's Hospital London, Glen Brice, Genetics Counsellor at South West Thames Regional Genetics Service, St George's University of London, Sahar Mansour, Consultant Geneticist at South West Thames Regional Genetics Service, St George's University of London, and Peter Mortimer, Professor of Dermatological Medicine, Cardiac and Vascular Services (Dermatology) St George's University of London</i> .....	p.4
<b>3. Living with lymphoedema: the experiences of children and their family</b> <i>Adapted from an original article by Christine Moffatt, CBE, Hon Professor in Nursing and Health Care Glasgow University, Director of the Centre for Research and Implementation of Clinical Practice (CRICP), and of the International Lymphoedema Framework (ILF) and Susie Murray, Researcher at the Centre for Research and Implementation of Clinical Practice, London</i> .....	p.9
<b>4. Services available for children and families living with lymphoedema</b> <i>Adapted from an original (unpublished) article by Dr Jacquelyne E Todd, Physiotherapist Consultant in Lymphoedema, Leeds Lymphoedema Service</i> .....	p.13
<b>5. Frequently asked questions</b> .....	p.17
<b>6. Syndromes and presentations</b> <i>Written by Gail Close, Clinical Manager, Haddenham Healthcare</i> .....	p.19
<b>7. References</b> .....	p.21
<b>8. Glossary</b> .....	p.22

# 1. Introduction

*This focus document will provide practitioners with an overview of childhood lymphoedema: the causes, presentations, and approaches to management.*

*Lymphoedema affecting children, teenagers and young adults is a rare chronic condition that has received very little attention due to poor medical knowledge. In most cases it is due to abnormalities in the development of the lymphatic system in the embryo, although it may develop as a result of cancer treatment or trauma. Lymphoedema due to Filariasis may also present in children. Lymphoedema is recognised by the World Health Organization (WHO) as a rare disease but also as an orphan one, meaning there is currently no cure.*

*The first description of congenital lymphoedema was given in 1891 by Milroy, with teenage lymphoedema described in 1898 by Meige. At that time, these works facilitated the differentiation between lymphoedema and other systemic forms of oedema. The recent identification of genes associated with hereditary lymphoedema has greatly enhanced the potential for the development of cures in the future.*

*The chronic nature of the condition impacts on many aspects of children's lives, particularly social, family and educational. Health care professionals working with children with lymphoedema will recognise these issues and the importance of improving the research basis of care and coordination of services.*

*This focus document is the initial step of our international strategy to define and improve the care of children with lymphoedema that firmly encompasses the views and needs of patients and their families.*

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## 2. Childhood lymphoedema: syndromes and presentations

Lymphoedema is chronic swelling, generally of the extremities, resulting from the failure of the lymphatic system to drain lymph fluid from the interstitial spaces<sup>1</sup>. Primary lymphoedema is caused by a developmental abnormality of the system and its prevalence in children, based on attendance at a single clinic, is estimated at 1.15 per 100,000 population<sup>2</sup>. Secondary lymphoedema is a consequence of removal or damage to lymph nodes (for example, after surgery), fibrosis of the nodes (post-radiotherapy) and injury or infection. Given that in the United Kingdom (UK), a physician is likely to see less than ten cases of lymphoedema in children per annum, diagnosis and thus rapid access to treatment and support is challenging<sup>3</sup>. (Box 1)

### Box 1: Obtaining an accurate diagnosis

“... the geneticist said there’s something not quite right... let’s wait and see what happens. The paediatrician said we will send you to an orthopaedic surgeon to have his feet reshaped... in the meantime, the plastic surgeon said ... this baby has lymphoedema...”

Parent<sup>4</sup>

### Common forms of childhood lymphoedema

Primary lymphoedema, occurring as a non-syndromic mendelian (inherited) condition, or as part of a syndromic disorder, is the most common type seen in children. In most cases, oedema will be present from birth, although may develop later in some cases.

While there are many forms of primary lymphoedema, the two main causes in children are Milroy disease and lymphoedema distichiasis. To ensure an accurate diagnosis, physicians will need to consider:

- The age of onset
- The location of the oedema
- Associated features
- Inheritance patterns
- Any underlying genetic cause

# Childhood lymphoedema: syndromes and presentations



**figure 1**  
 Demonstration of positive Stemmer sign, the inability to pick up a fold of skin at the base of the second toe, indicates thickening of the skin and is useful in differentiating lymphoedema from other forms of oedema. Also known as Kaposi-Stemmer sign.

## Diagnosing lymphoedema

Diagnosis is based primarily on clinical presentation, although it should be distinguished from other conditions with overlapping phenotypes in order to instigate the appropriate treatment. Physicians need to be aware that primary lymphoedema can coexist with secondary causes; finding a secondary cause does not exclude a primary weakness of the lymphatic system.

A range of clinical features present in lymphoedema including:

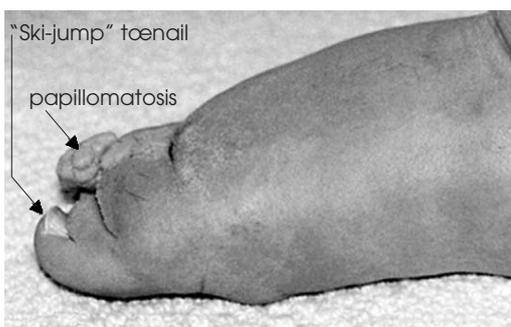
- Cutaneous and subcutaneous thickening
- Pitting oedema (which does not resolve significantly with elevation)
- Positive Kaposi-Stemmer sign<sup>5</sup> (figure 1)
- Fibrosis
- Hyperkeratosis
- Papillomatosis
- Lymphangiectasia

When there is systemic involvement, ascites, pleural effusion, and intestinal lymphangiectasia can occur.

Lymphoscintigraphy is the 'gold standard' for lymph system investigation, with functional imaging and lymphatic-specific markers as adjuncts.

## Milroy disease

Milroy disease, a congenital onset lymphoedema, classically affects the lower half of the legs<sup>6</sup>. Clinical findings include upslanting toenails (figure 2), prominent large calibre veins in the legs (figure 3), and in males, hydroceles in 30% of cases.



**figure 2**  
 Adult patient with Milroy disease showing pedal oedema, 'ski jump' nails and papillomatosis due to congenital lymphatic abnormalities.



**figure 3**  
 Prominent veins, commonly seen in Milroy disease.

# Childhood lymphoedema: syndromes and presentations

## Lymphoedema distichiasis

This rare condition is the association of primary, pubertal or post-pubertal onset lymphoedema with aberrant or extra eyelashes (*figure 4*). Also noted are cleft palate, varicose veins and venous abnormality, and congenital heart disease.

Lymphoedema is also associated with a number of syndromic conditions such as

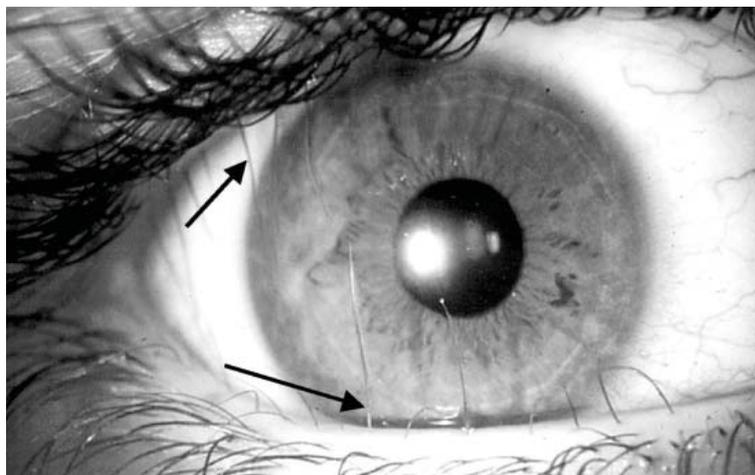
- Turner
- Noonan
- Prader-Willi
- CHARGE (Coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, ear abnormalities/deafness)
- Klippel-Trénaunay Syndrome

*(see section 6 for full presentations of these syndromes)*

Other causes and associations are outlined in box 2.

## Complications

The number, nature and severity of complications is dependent on the degree of oedema. Cellulitis and lymphangitis are common. Cellulitis can lead to acute illness and further damage to the lymphatic system. Disturbed cellular trafficking resulting from abnormal lymph drainage can impair the immune system; a local infection can rapidly deteriorate into septicaemia.



**figure 4**  
*Distichiasis on upper and lower lids. Note presence of more than one row of normal lashes but anterior to, and clearly separate from the abnormal lashes (arrowed) growing from the Meibomian gland*

# Childhood lymphoedema: syndromes and presentations

## Box 2: Other causes of and associations with lymphoedema

### Vascular malformation

*Segmental lymphoedema:* A recognised phenotype, where the lymphoedema affects one or more body segments and adjacent structures (genitalia, face, conjunctiva). No systemic involvement (*figure 5*).

*Generalised lymphatic dysplasia (types I & II):* Systemic involvement with multi-segmental effects (type I), or generalised, affecting the whole body (type II) (*figure 6*).

## Management

Unfortunately, lymphoedema is a life-long condition and requires multiprofessional input at all stages. It should be made clear to the child and family that the emphasis is on management rather than cure. While there is no firm guidance on the treatment and prophylaxis, a document pertaining to adults can be adapted for children:

[www.lymphoedema.org/lsn](http://www.lymphoedema.org/lsn)

The lymphoedema therapist is key to the delivery and coordination of

care. Basic management principles include:

- Reduction of swelling and improvement of shape
- Prevention and treatment of infection
- Treatment of skin problems such as papillomatosis and lymphorrhoea
- Pain management
- Psychosocial intervention
- Physical therapy; to control lymph formation and improve drainage (exercise, external compression, manual lymphatic drainage)

Clearly an early and accurate diagnosis is essential to instigate appropriate management. (Box 3 summarises considerations for diagnosis.)

**figure 5**

*Unisegmental oedema in a child*



**figure 6**

*Widespread lymphatic dysplasia with four limb oedema and ascites.*

## Childhood lymphoedema: syndromes and presentations

<b>Box 3: Considerations for diagnosis</b>	
<b>Primary or secondary; rule out secondary cause</b>	<ul style="list-style-type: none"> <li>■ Injury</li> <li>■ Drugs</li> <li>■ Overseas travel (exotic infection)</li> <li>■ Surgery</li> <li>■ Neurological conditions</li> <li>■ Obstruction or malignancy</li> </ul>
<b>Age of onset</b>	<ul style="list-style-type: none"> <li>■ Congenital</li> <li>■ Pubertal</li> <li>■ Other</li> </ul>
<b>Family history?</b>	<ul style="list-style-type: none"> <li>■ History of lymphoedema and associated issues (such as hydrocele, cellulitis, veins)</li> </ul>
<b>Any associated features suggesting a recognisable syndrome?</b>	<ul style="list-style-type: none"> <li>■ Dysmorphic features</li> <li>■ Learning difficulties</li> </ul> <p><i>If present, consider genetics referral</i></p>
<b>Investigations</b>	<ul style="list-style-type: none"> <li>■ Albumin</li> <li>■ Markers of immune function</li> <li>■ B-type natriuretic peptide (BNP)</li> <li>■ Renal function tests</li> <li>■ Lymphoscintigraphy</li> <li>■ Abdominal ultrasound</li> <li>■ Venous duplex ultrasound</li> <li>■ MRI scan, X-Ray</li> </ul>
<b>Genetic testing</b>	<ul style="list-style-type: none"> <li>■ Mutation analysis</li> <li>■ Standard karyotyping</li> <li>■ Comparative genome hybridisation (CGH) if dysmorphic or has learning disabilities</li> </ul>
<b>Management</b>	<ul style="list-style-type: none"> <li>■ Referral to lymphoedema therapist</li> <li>■ Compression garments and exercise</li> </ul> <p><i>Infection control</i></p>

This section is adapted from an original article<sup>7</sup>.

# 3. Living with lymphoedema: the experiences of children and their family

While relatively little is known about the effects of lymphoedema on children and their families, anecdotal evidence suggests that one of the biggest challenges faced is accessing treatment as services are sparse.

Moffatt and Murray<sup>4</sup> studied 20 families to explore the experience of living with lymphoedema on parents, children and adolescents.

The stories that unfolded showed that at every stage, from realising that the child had a problem to accessing treatment and on-going management services, was fraught with difficulty and anxiety inducing; for example, it was not uncommon for professionals to suggest that the parent(s) had contributed to the development of lymphoedema, asking the mother for example, if she had smoked or drunk during pregnancy (Box 4).

## Diagnosis

Obtaining a correct diagnosis can take some considerable time. For example, of the 20 children included in the study:

- 4 were diagnosed before birth
- 8 were diagnosed at birth
- 8 were diagnosed in early childhood

Families often see a plethora of professionals who either do not have the knowledge or skill to diagnose the problem, or who attribute it to another cause.

## Box 4: Families' experience of professionals<sup>4</sup>

- 'Live with it'
- Lack of local services
- General lack of knowledge and interest both at primary and secondary care level
- Poor interpersonal skills
- Arrogant attitudes towards other professionals in different disciplines

## Living with lymphoedema: the experiences of children and their family

### Accessing expert advice

Accessing information on the condition is difficult and where it is found, relates mainly to adults. Professionals are not always able to predict the course of a child's disease, so it is assumed that it would be that of an adult (Box 5).

No single source of clinical or practical advice exists (for example, where to obtain extra products), which can lead to conflicting information being given. Parents may have to take on the role of educator to professionals in order to ensure that their child receives the appropriate treatment.

However, once expert advice, for example at a centre of excellence, was found, staff were able to reassure parents and provide sound clinical information and information on organisations such as the Lymphoedema Support Network<sup>8</sup> (LSN) and local support groups. Unfortunately, many such centres are not local to the family, so attending appointments can require a reorganisation of the whole family, for example, siblings having to be left with friends. Such disruption to family life is time consuming, expensive in terms of travel and accommodation and may lead to feelings of sibling exclusion.

Disappointingly, once treatment for example, manual lymphatic drainage, and information has been accessed at the specialist centres, parents often find that these are not available (or were not funded) locally, so resort to paying for this themselves. In addition, 'maintenance' services such as podiatry were denied unless a referral was given from the specialist centres.

### Box 5: Parent experiences

...he read the explanation from a medical textbook. He knew nothing about it. ...my husband and I were quite devastated... because it sounded so bad. ”

Parent<sup>4</sup>

### The burden to parents

The challenges that parents face cannot be underestimated. Much centres on being an advocate for their child as they attempt to overcome the bureaucracy, lack of information and attitudes of professionals (Box 6).

Ensuring that schools are prepared to cope with the child is hard as many are inflexible in addressing both simple and specific requirements; for example, staff not helping the child to reapply hosiery after sport. Bullying is also an issue, as is the attitude of staff, some suggesting that mainstream schooling is not appropriate.

### Box 6: Issues facing parents

- Searching for a diagnosis
- Finding information
- Pushing for expert care and appropriate service; for example, podiatry
- Lack of continuity in services
- Advocate for child's needs
- Learning and managing prescribed treatment
- Ensuring an adequate supply of pressure garments, shoes and bandages (lack of funding or coordinated ordering and delivery)
- Understanding and managing physical and psychosocial needs of affected child and siblings; for example, the child may be bullied, unhappy with hosiery/shoes, siblings may feel excluded and/or jealous
- Allowing children 'normality' despite the problems that the condition brings. This is an issue at all ages, but particularly during adolescence as they strive towards independence

### The child's experience

Young children who have never known life without lymphoedema are generally accepting of it and its attendant problems, particularly if it restricted an activity they enjoyed, such as sport. Most had an understanding of the condition and the long-term implications, but are often hopeful for a cure.

Children find their own mechanisms for coping with the frustration they felt;

*“... you just have to kick and punch the pillows...if you have a hill near you, ... run down it screaming and waving your arms like a maniac and when you get to the bottom, you're feeling much better...”*

Sharing their experiences with other children is also enormously helpful, diminishing feelings of isolation.

Lymphoedema has a much greater impact upon adolescents, particularly in terms of body image and peer-group inclusion; coping mechanisms included dressing in such a way as to hide the swelling or not telling others they had it. Concordance is often difficult, after all, what teenager wants to wear a compression garment? Again, meeting with other teenagers with the same condition was helpful.

## Living with lymphoedema: the experiences of children and their family

### **Towards a proactive and seamless service**

While the experiences of the families caused enormous distress and anxiety, and particularly where lymphoedema was familial, it was notable that these emotions were then used positively – parents became strong advocates for their children. Their knowledge and experiences should be acknowledged by professionals and used to develop management strategies for each individual family.

Moffatt and Murray<sup>4</sup> suggest a number of steps that can be taken to facilitate a proactive and seamless service for them and their family:

- An easier path to diagnosis with professionals able to access appropriate funding and other professionals
- Professionals (medical and educational) work with parents to meet the best interests of the child. Those in primary care to recognise the knowledge and experience of specialists and act accordingly
- Continuity of care, particularly if a local service ceases to exist and when moving from child to adult services
- Non-specialist practitioners to be more aware of lymphoedema and its management and access to their services (such as podiatry) freely and simply available
- Better access to information on the condition and management options at levels appropriate for the parents and the child and/or adolescent to strengthen their understanding
- Contact and interaction with other families of children with lymphoedema
- Appointment times and venues to meet the needs of professionals and families
- Providing information about benefits (such as travelling expenses)
- Ensuring regular fitting and supply of hosiery and shoes (with potential to purchase extra where required). Developing these so that they are acceptable to children and adolescents

Further information can be obtained from:

**[www.lympho.org](http://www.lympho.org)**

**[www.lymphoedema.org/lsn](http://www.lymphoedema.org/lsn)**

# 4. Services available for children and families living with lymphoedema

Dedicated services for children with lymphoedema are rare, with no clear guidelines or standards for their provision. Currently, guidance such as that contained in the Department of Health (DH) and Department for Children, Schools and Families (DCSF) guidelines<sup>9,10</sup> outlines core principles and national standards to ensure that the child's spectrum of needs is met (Box 7). Clinically, treatment of childhood lymphoedema is based on adult guidelines<sup>11,12</sup>. Ideally, the recommendations within these documents, along with the recommendations of Moffatt and Murray<sup>4</sup>, can dovetail into a dedicated framework for children's lymphoedema management and services.

## **Box 7: General principles of the National Service Framework for Children, Young People and Maternity Services<sup>9</sup>**

- Coordinated multi-agency care
- Empowering children and young people to take responsibility for their actions and make informed choices about their health
- Provision of support, education and information
- Accessible services that are responsive to the needs of children and families
- Staff share a common core of skills, knowledge and competencies
- Services are age-appropriate and address needs associated with the condition in other aspects of life
- Access to high-quality treatment that addresses all aspects of children's welfare
- Care delivered in appropriate settings

## Services available for children and families living with lymphoedema

Due to the congenital nature of primary lymphoedema, diagnosis and thus referral for treatment can take place at any time between the antenatal period and puberty. Naturally, this means that the needs of the child and the parents in terms of professional and clinical input, psychological care and resources, will be varied.

Service providers also need to consider the transition from child to adult services, which may not necessarily be age dependent. Current DH guidance<sup>9</sup> suggests that children and young people are those below 19 years of age, although this may vary according to the statutory obligations of individual agencies. Where children have access to small, expert units, the transition is generally seamless as they see the same professionals. This can be facilitated by the same clinician continuing the lead role in provision of care and providing young adult specific clinics for social interaction, education and information exchange and to reduce feelings of isolation.

### Multidisciplinary approach to care delivery

A team-based approach is required to fully meet the needs of the child. This should comprise:

- **A specialist/advanced lymphoedema practitioner**  
Specialists have the experience and clinical skills to address the physical, functional and psychosocial needs of the child and family; coordinate care; communicate with multiple agencies (for example, the child's school).
- **A paediatrician and a geneticist**  
The latter will identify inherited forms of lymphoedema and provide counselling and information as to the trajectory of the problem; the paediatrician can offer specific medical advice on the problems associated with lymphoedema such as cellulitis, and refer to medical colleagues as required.
- **Clinical psychologist**
- **Play therapist**
- **Others**  
Dermatologist, physiotherapist, occupational therapist, surgical fitter, dietician, surgeon.

# Services available for children and families living with lymphoedema

Lymphoedema services will have to comply with national guidelines and local protocols to determine where and how treatment can be provided. The environment should be child-orientated and if in an existing children's unit, provide space and resources for lymphoedema services, a separate waiting area, and planned sessions exclusively for these children with appropriate resources, facilities and child-specific information.

Developing a dedicated service for children and young people with lymphoedema within existing local units can be difficult, and may not necessarily afford the opportunity to meet other children with lymphoedema. A regional facility would afford better opportunities and offer standardised high-quality care to a larger number of children. However, communication between the centre of excellence and local providers needs to be strong, contemporaneous and rapid to ensure continuity of care and help reduce family anxiety. Most importantly, where appropriate and with the relevant permissions, pertinent information must be shared between agencies. Where children are at risk of poor outcome as a result of additional health, education or social needs, the Common Assessment Framework for children and young people: practitioner's guide<sup>13</sup> provides a standardised assessment approach.

As previously noted, best practice for the management of lymphoedema in children and young adults should incorporate a number of existing guidelines, policies and recommendations. Central to this should be the following standards:

### **Identification of people at risk of or with lymphoedema**

This can be compromised by lack of public and professional awareness. Providing specific information may help the profile of the condition and facilitate children and families to access high-quality treatment and support.

### **Empowerment**

The needs of the children and their families may change quickly, so in order to adapt plans of care and access to services, regular appointments should be available. Access to advice, support and information should also be available through for example, the telephone and internet.

## Services available for children and families living with lymphoedema

### ***Provision of lymphoedema services that deliver high-quality clinical care that is subject to continuous improvement and that integrates community, hospital and hospice-based services***

To achieve its maximum physical and functional potential and to minimise distress, the child requires high-quality clinical care. This care needs to be integrated into the family lifestyle, avoiding disruption to normal life and be congruent with the child's needs where possible to avoid non-concordance.

### ***Provision of high-quality clinical care for people with cellulitis/erysipelas***

Adult protocols are used but the dosage adjusted according to the size of the child.

### ***Provision of compression garments for people with lymphoedema***

Children require different functional and cosmetic properties from compression garments. Types of activities undertaken along with normal growth and shape-change mean that they will require regular fittings and more than two sets every six months.

### ***Provision of multi-agency health and social care***

This is central to national guidance on children's services. Practitioners must be aware of such services, and how to facilitate referral.

### **Conclusion**

Children's services should be offered through a network of regional centres that deliver standardised, high-quality care, tailored to the individual child and family and that is responsive to their needs.

Further information can be obtained from:

**[www.lymphoedemaleeds.co.uk](http://www.lymphoedemaleeds.co.uk)**

**[www.lymphoedema.org/ltn/](http://www.lymphoedema.org/ltn/)  
(child and teenager fact sheets)**

# 5. Frequently asked questions

Staff involved in the care of children and their families should be prepared to answer many questions. Naturally, many will relate to the effect of lymphoedema on their lives, so answers will be unique to that family. However, the most frequently asked questions are presented here.

- Are there many children with lymphoedema?
- As our first child has lymphoedema, how likely is it that any other children we have will also have the condition?
- Why are specialist services so far from our local area?
- Why are there so few experts in the field of lymphoedema causing us to have to wait a long time for diagnosis?
- I understand that lymphoscintigraphy may help us understand the cause of my child's lymphoedema but will it help in what treatment he will receive?
- Will my child have to wear bandages/hosiery for ever?
- How do you deal with itching caused by bandages – it is distressing to the child?
- Putting the stockings on my child is difficult. Is there any practical help for this?
- How can we get supplies of hosiery when our child needs it?
- Can we pay for extra hosiery if our child needs it?
- Our daughter is unable to buy shoes in the High Street so they are made at the hospital. Can she have more than one pair at a time?

# Frequently asked questions

- Can the shoes provided by the hospital be more fashionable?
- Where can I buy clothes that will fit my child and that are age-appropriate?
- Why are children not offered preventative MLD?
- Can we get funding for MLD?
- What measures can I take to help prevent infections for my child?
- How do I recognise cellulitis?
- How can I access treatment for my child's recurrent verrucas?
- How can we ensure schools understand our child's condition?
- How can we help our child who is being bullied at school because of the hosiery/shoes that he wears?
- Are there any support groups for parents of children with lymphoedema?
- Are there websites that could tell our child more about lymphoedema?

# 6. Syndromes and presentations

## Milroy Disease

Termed as congenital hereditary lymphoedema of the legs

- Present in one or both legs at birth
- Family history of similar swelling
- Genetically linked to a breakage in the VEGFR3 gene

## Klippel-Trénaunay Syndrome (figures 7&8)

Characterised by some or all of the following:

- Port wine stain or birthmark (cutaneous capillary malformations)
- Soft tissue and bony hypertrophy
- Venous malformations
- Lymphatic abnormalities
- Usually limited to one limb but may occur in multiple limbs and/or head and trunk area

[www.k-t.org](http://www.k-t.org)

### figures 7&8

18 year-old girl with Klippel-Trénaunay recently diagnosed - prior to treatment



## Turner's Syndrome

*Affects females only*

Characterised by some or all of the following:

- Broad chest
- Widely spaced nipples
- Droopy eyelids
- Low hair line
- Low set ears
- Spoon shaped, or small, or hyperconvex nails
- Short fingers and fourth toe
- Webbed neck
- High arched palate
- Short stature
- Cubitus valgus (turning in of the elbows)
- Otitis media
- Hearing problems
- Myopia
- Pigmented naevi
- Infertility
- High blood pressure
- Kidney and urinary tract problems
- Coarctation (narrowing of the aorta)
- Thyroid problems
- Small lower jaw
- Osteoporosis
- Diabetes mellitus
- Behavioural problems
- Learning difficulties

[www.tss.org.uk](http://www.tss.org.uk)

# Syndromes and presentations

## Noonan's Syndrome

Characterised by some or all of the following:

- Drooping eyelids
- Large, downward slanting eyes
- Widely spaced eyes
- Strikingly blue or blue/green eyes
- Low set ears
- Flat bridge to the nose
- Wide forehead
- Round shouldered look
- Low neck hair line
- Short stature
- Congenital heart defects
- Breast bone deformities with a sunken chest
- Clotting problems
- Myopia, squint or astigmatism
- Weak muscle in the mouth
- Delayed speech development
- Delayed onset of puberty

[www.noonansyndrome.org](http://www.noonansyndrome.org)

## Prader-Willi (figures 9&10)

Characterised by some or all of the following:

- Hypotonia
- Hypogonadism
- Obesity and hyperphagia
- Learning difficulties
- Short stature
- Small hands and feet
- Somnolence
- Poor emotional and social development
- Almond shaped eyes
- Narrow forehead
- Down turned mouth with triangular shaped upper lip
- Poor coordination and balance
- Curvature of the spine

[www.praderwillisyndrome.org.uk](http://www.praderwillisyndrome.org.uk)



◀ **figure 9**  
20 year-old man with Prader-Willi



▲ **figure 10**  
21 year-old man with Prader-Willi with wounds

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# 8. Glossary

## **CHARGE**

Coloboma, heart defect, atresia choanae, retarded growth and development, genital hypoplasia, ear abnormalities/deafness)

## **Complete decongestive therapy/physiotherapy (CDT/P)**

Comprises 4 elements of therapy to manage lymphoedema:

- Compression (bandages or garments)
- Manual lymphatic drainage
- Exercises
- Skin care

## **Fibrosis**

Thickening of the skin and underlying structures due to the inefficient drainage and build-up of protein-rich lymph fluid

## **Hyperkeratosis**

Thickening of the skin caused by excessive production of keratin

## **Kaposi-Stemmer sign**

Inability to pinch a fold of skin at the base of the second toe

## **Lymphangiectasia**

Vesicles resulting from engorged lymphatic vessels

## **Lymphoedema**

Swelling (generally of limbs) due to the accumulation of fluid in the interstitial tissue as a result of lymphatic system failure

## **Lymphorrhea**

Leakage of lymph fluid from vesicles

## **Manual lymphatic drainage**

Massage-like manipulation of the affected area following the direction of lymph flow to stimulate lymph movement

## **Papillomatosis**

Pouching of pockets of skin between fibrosed areas

## **Primary lymphoedema**

Lymphoedema caused by a developmental abnormality of the lymphatic system

## **Secondary lymphoedema**

Lymphoedema resulting from removal or damage to lymph nodes (for example, after surgery), fibrosis of the nodes (post-radiotherapy) and injury or infection



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### Designed by

Couleur Café  
*Monistrol sur Loire  
France*

### Produced by

Concept Design,  
*Berkhamsted,  
England*

## Acknowledgements

The International Lymphoedema Framework would like to thank:

- The children and their families who have kindly given permission for the reproduction of the images in the document
- The clinicians who have contributed to and reviewed the document: Isabelle Quéré, Christine Moffatt, Jacquelyne Todd, Fiona Connell, Glen Brice, Sahar Mansour, Peter Mortimer, Gail Close, Susie Murray
- The editor and publisher of the Journal of Lymphoedema for their permission to use the Connell, F., Brice, G., Mansour, S. et al (2009) article in section 2



# The ILF Objective :

## To improve the management of lymphoedema and related disorders worldwide

- To increase **awareness** by raising the profile of lymphoedema.
- To increase **knowledge** about lymphoedema by initiating and/or contributing to **Research Programmes**.
- To **disseminate** this knowledge by implementing an **international, not-for-profit, publications strategy**.
- To increase **understanding** of lymphoedema and its management by creating and/or contributing to the development of **Education Programmes**.
- To provide a cross cultural networking platform through an **Annual International Event** where all stakeholders will have the opportunity to contribute and influence the ILF agenda.
- To promote and document Best Practice with the development of an **International Minimum Dataset**.
- To facilitate and/or contribute to better **access to treatment** for patients worldwide.
- To promote and **support initiatives** whose goals are to improve the national/regional/local management of lymphoedema anywhere in the world.
- To help the Healthcare Industry understand the **real needs** of patients and practitioners, and develop and evaluate improved diagnostic tools and treatments.

