Common skin problems in children
Part 4: birthmarks

Although most birthmarks are small and harmless, they can sometimes be large and cause cosmetic or functional impairment. Others can indicate an underlying abnormality, and a few have a malignant potential. Some birthmarks can be removed, but others remain a challenge to treat even with present surgical and laser techniques.

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To the layperson, the word 'birthmark' is a term that indicates a permanent skin lesion that a child is born with. However, not all congenital lesions are permanent, and not all permanent lesions are present at birth. From a medical viewpoint, birthmarks encompass a wide variety of cutaneous lesions, including malformations, neoplasms and hamartomas. The cells of these lesions include melanocytes, vascular elements, keratinocytes and skin appendages such as sebaceous glands, all of which occur normally in the skin.

This article, the last in the series of articles on common skin problems in children, reviews the most common types of birthmarks occurring in childhood.

Pigmented naevi

Melanocytic naevi

Most melanocytic naevi (often known as 'moles') are not present at birth, but they can be congenital, occurring in about one in 100 newborns (Figure 1). Usually melanocytic naevi are small (less than 2 cm in diameter), but rarely they may be very large (20 cm or greater in diameter).

Melanocytic lesions over 20 cm in diameter are termed 'giant' (Figure 2). They are invariably present at birth, occurring in one in 500,000 births, and are often covered in hair. They may be seen in a garment distribution on the trunk and may be surrounded by numerous satellite lesions. Classically, this type of lesion is associated with early onset melanoma.

Nearly all children will develop melanocytic naevi after the age of 2 years; they can appear at any time up to the age of about the mid-20s. Most of these naevi are completely harmless and do not require treatment. The number of these lesions is determined by a combination of genetic

IN SUMMARY

- **'Birthmark' is a lay term that has many medical equivalents, including neoplasms, hamartomas and malformations; not all are present at birth.**
- **Although most haemangiomas are uncomplicated, requiring no intervention, patients with facial haemangiomas need to be carefully observed; even a modest increase in size of these lesions can lead to substantial problems.**
- **The risk of melanoma in children is low; most melanocytic naevi are removed for cosmetic reasons, not malignancy.**
- **Children with capillary malformations (port wine stains) may be treated successfully with laser therapy; they should be referred for assessment by the age of 12 months.**
- **Naevus sebaceous is the most common epidermal naevus and one of the few with a malignant potential.**
- **Most small birthmarks can be removed; the decision to remove birthmarks should, in most cases, involve the child.**

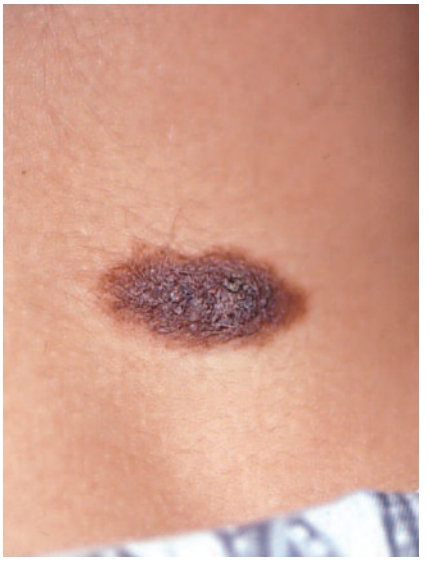


Figure 1. Congenital melanocytic naevus.

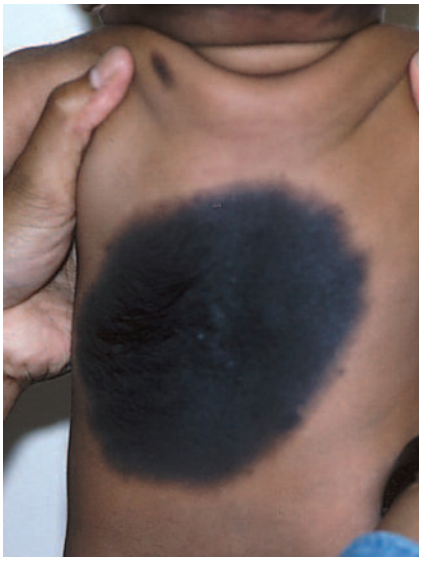


Figure 2. Giant melanocytic naevus.

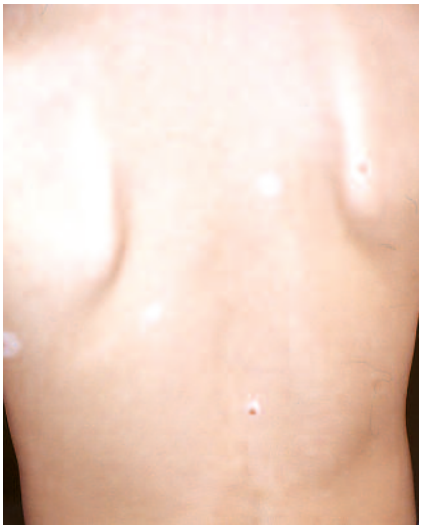


Figure 3. Halo naevus; a white halo may appear around a melanocytic naevus.

potential and degree of sun exposure. Although they are most common on sun exposed sites, melanocytic naevi can occur anywhere on the body. The appearance is very variable. Colour varies from pink to brown to black, the latter being seen particularly in dark skinned individuals. Naevi on the scalp often have a pale centre. The lesions may be associated with a variable amount of hair. The shape is usually round to ovoid, but it is not uncommon for the edge to be somewhat irregular.

It is not always appreciated that melanocytic naevi have a life cycle. It is normal for them to change slowly with time (over years) in size and colour, and eventually they involute so that elderly persons have very few. At puberty they often darken, enlarge and may become hairy, and it is not uncommon for these normal changes to cause concern about melanoma.

Certain events may complicate all pigmented naevi. Occasionally they may become itchy or swollen. In some cases, a white halo may appear around the naevus (halo naevus; Figure 3). In atopic children, dermatitis may localise around naevi. Pedunculated lesions may strangulate, making them look suddenly and temporarily black. All these changes are benign.

The risk of melanoma in children with small melanocytic naevi is negligible; melanoma in prepubertal children is very rare. In those with large congenital naevi, melanoma may occur before puberty. In at least half the cases of melanoma occurring in patients with giant melanocytic naevi, the malignancy is found in areas other than the naevus and, therefore, removing the naevus does not completely remove the melanoma risk.

Melanoma risk is correlated with numbers of naevi and family history of melanoma. Some families have a history of numerous, unusual looking naevi that are associated with multiple melanomas. This is known as the 'dysplastic naevus syndrome' and is quite uncommon. Many patients with multiple naevi never have a melanoma.

In children the strongest indicator for removal of most melanocytic naevi, particularly small ones, is not concern about their malignant potential, but cosmetic embarrassment. Surgical removal of cosmetically distressing lesions can be contemplated before the child starts school. Sometimes the appearance of the lesion can be improved simply by epilating hair, if present; this may be achieved with hair removal laser therapy. Response to other laser therapy is poor.

Removal of large lesions is rarely easy and may involve numerous complex surgical procedures. Curettage in the neonatal period has been described recently as a treatment option. Outcomes are variable but sometimes very good.

Giant melanocytic naevi may also be treated with dermabrasion and laser therapy, although the latter tends to be the least effective modality. These cosmetic procedures still leave a cancer risk, and the cosmetic effect may not be lasting. Referral to a dermatologist is recommended for any newborns with such giant lesions.

Mongolian spot

Mongolian spot is a very common congenital lesion in Asian babies, but it is seen occasionally in Caucasians. Typically, it is a blue-grey macule, occurring most often on the lower back (Figure 4), although it can be found on the limbs. Some are quite large. Spontaneous resolution usually occurs during the first decade of life.

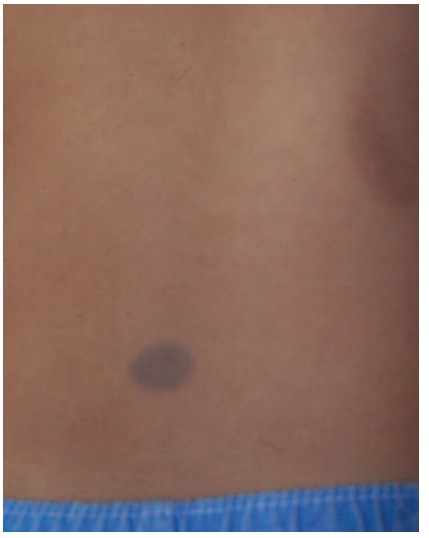


Figure 4. Typical appearance of a Mongolian spot.

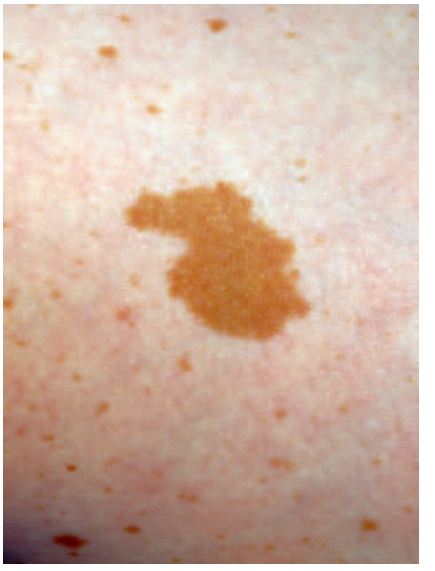


Figure 5 (above). Café au lait macule.

Café au lait macule

Although most doctors think of neurofibromatosis when they see the flat, well-demarcated, light brown lesions of café au lait macules, these lesions are in fact common as solitary birthmarks and are both benign and without significance (Figure 5). They are permanent and darken in summer with sun exposure. Neurofibromatosis needs only be considered when six or more lesions are present, particularly if there is freckling in the axillae and groin.

Café au lait macules can be lightened with laser treatment but have a tendency to recur.

Epidermal naevi

Epidermal naevi are the most uncommon and heterogenous group of naevi. The lesions are composed of any element present in the epidermis and dermis; most of them are hamartomas.

Some epidermal naevi are quite extensive, occurring over large areas of the body in linear whorls and streaks. In these cases, genetic mosaicism is believed to be involved. It is not surprising, therefore, that other abnormalities may be associated, particularly of the skeleton, eyes and central nervous system. This association is known as the 'epidermal naevus syndrome' and is highly variable. Children with large epidermal naevi (Figure 6) should be referred to a paediatrician for evaluation.

Epidermal naevi are not always present at birth. They may occur for the first time in early childhood and then extend for several years. It is not uncommon for them to be linear, extending all the way down a limb (Figure 7). Like melanocytic naevi, they not uncommonly enlarge and

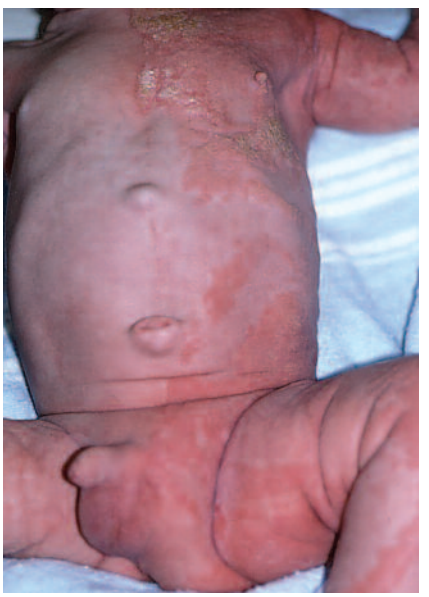


Figure 6. Large epidermal naevus; patients with such lesions should be referred to a paediatrician.

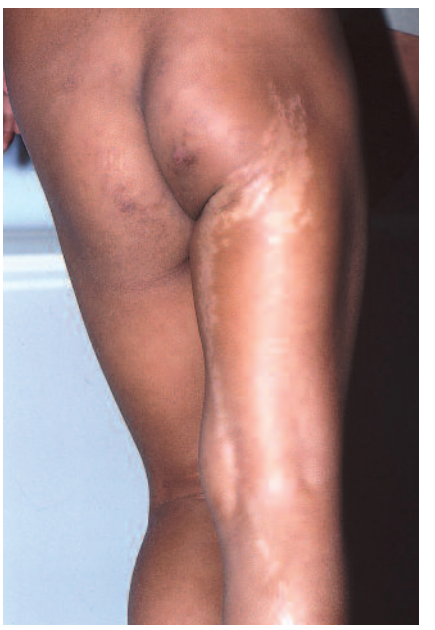


Figure 7. Epidermal naevi may be linear, extending down a limb.

become more problematic at puberty, mainly because of cosmetic and functional concerns. Malignancy is rarely a complication, except in patients with naevus sebaceous.

The most common member of this group is the naevus sebaceous (Figure 8), consisting predominantly of sebaceous glands. It usually occurs on the head and neck, and is present at birth. It appears as a hairless, orange-yellow plaque. Unlike most other epidermal naevi, this particular lesion has a small malignant potential and may be complicated by carcinoma, most often basal cell carcinoma, in late teenage to adult life. Because of this, it may be wise to remove these lesions prophylactically by the time the patient is 15 years old; they are rarely large or difficult to excise.

Naevi composed of keratinocytes (verrucous epidermal naevi) have the appearance of warty linear streaks (Figure 9) and may be mistaken for warts or even lichenified dermatitis.

If localised, the best treatment for epidermal naevi is complete full-thickness excision. Laser therapy is helpful but recurrence after this sort of treatment is usual. In most cases, these lesions are benign and only a cosmetic problem. In some locations, however, such as the genital area or fingers, they may become a functional problem. These naevi are sufficiently unusual to warrant referral of affected patients to a dermatologist in most cases.

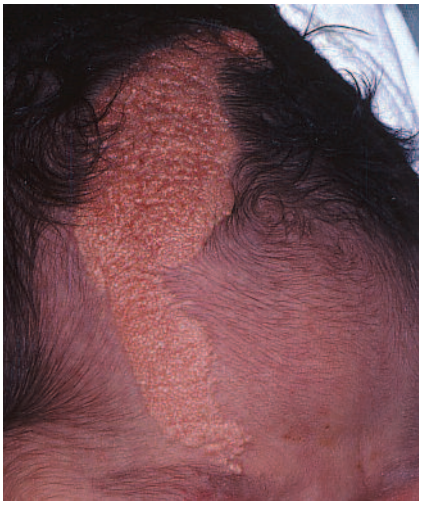


Figure 8. Naevus sebaceous. Such lesions have a small malignant potential.

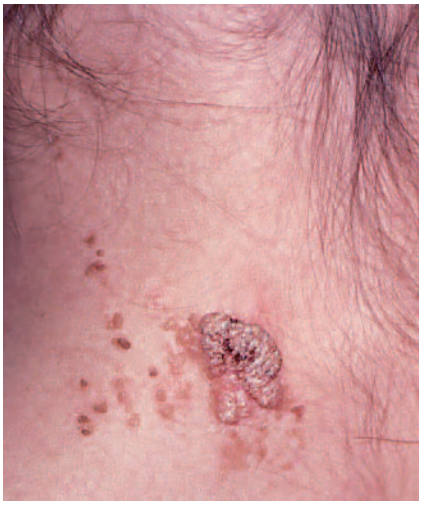


Figure 9. Warty appearance of verrucous epidermal naevus.

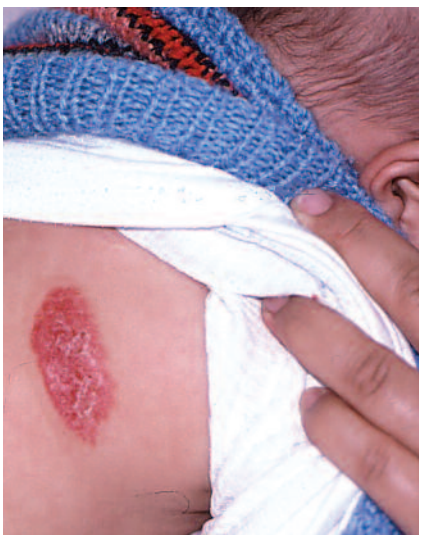
Vascular birthmarks

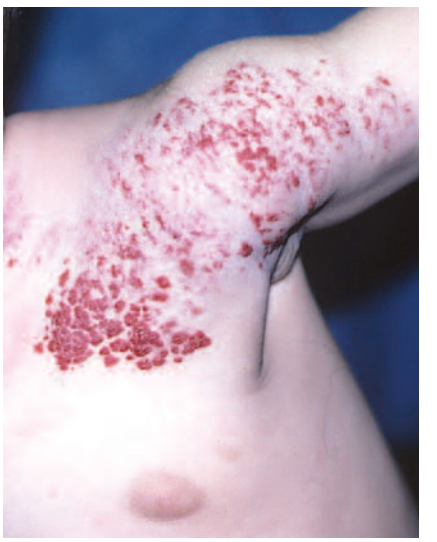
Haemangioma of infancy

Haemangioma of infancy (previously called capillary haemangioma and strawberry naevus; Figures 10a and b) is a common neoplasm affecting 10% of neonates. It is more common in girls and premature babies. Although not usually present at birth, it appears within the first month of life, often initially as an area of pallor, erythema or telangiectasia. There is then a period of growth for up to 20 weeks, rarely reaching very large proportions. After stabilising, the lesion slowly regresses, resolving substantially by the time the child is of school age and completely by the age of 9 years.

Many of these lesions are superficial, but they may have a deep component. The characteristic appearance is of a bright red nodule but the deep part appears bluish and is obviously below the surface of the skin. Sometimes the entire lesion is deep, making it somewhat difficult to distinguish from a vascular malformation (see below). During regression, grey areas appear on the surface and are a good prognostic sign. The superficial portion usually resolves before the deep one does. In 20% of cases there is more than one lesion.

Most of these lesions are uncomplicated and never require any form of intervention. When complications do occur, the most common is ulceration. This occurs most often under the nappy





Figures 10a and b. Haemangioma of infancy. This common neoplasm affects 10% of neonates.

and on the lip. It is important to be aware of this as a complication because children so affected are sometimes reported as being cases of child abuse. Ulceration is usually treated conservatively with occlusive dressings; however, persistently ulcerated lesions may benefit from surgical or pulsed dye laser treatment. (The use of pulsed dye laser for haemangiomas is controversial, and is useful only for flat, superficial lesions.) Referral to a dermatologist is recommended in cases such as these.

Other complications are unusual. Bleeding and infection occur rarely, and in a child with dermatitis, this may localise to the lesion. Parents often say that the worst thing about having a child with a haemangioma, particularly on a visible area such as the head and neck, is the comments they receive from complete strangers in public places, particularly supermarkets. Parents often need support and encouragement to develop resilience to this intrusion on their privacy.

Most of these lesions do not cause a problem and, because of their natural history, no treatment is recommended; however, beware the following situations:

- facial lesions that may become disfiguring if they enlarge
- lesions that interfere with an orifice (ear, nose, genitals, mouth)
- lesions that may occlude an eye (which may cause blindness)
- rapidly growing large lesions
- large, flat facial lesions around the mouth (these are associated with the very dangerous laryngeal haemangioma that can cause airway obstruction).

In these cases, therapy with high dose oral corticosteroids to halt progression and speed of involution should be started as early as possible, and urgent referral to a paediatrician or dermatologist is essential. Any baby with such a lesion should be closely observed.

Large haemangiomas of infancy may leave residual stretched tissue and telangiectasia, which may require later surgical intervention and pulsed dye laser therapy.

Naevus flammeus

Naevus flammeus, also known as a 'stork mark' is seen in 50% of newborns. It is found on the glabella, upper eyelids and nuchal area as irregular red macules that become more obvious when the infant cries.

When these lesions occur on the face they invariably resolve by 12 months of age. However, those occurring on the nuchal area persist into adult life in many cases, and are found in 10 to 20% of adults.

Capillary malformation

Capillary malformation (port wine stain; Figure 11) is encountered in 1 in 1000 births. Although present at birth, it is not always obvious and may not be diagnosed for several months.

The lesion consists of excess superficial capillaries that were formed early in fetal life. Because of this, the lesion may not be confined to the skin; the cutaneous lesion may be the 'tip of the iceberg', with underlying abnormalities of vessel, soft tissue, nerve and bone. Unlike haemangiomas, these lesions are permanent, and do not undergo a growth and resolution phase. Generally, they tend to deteriorate with age.

When found on the face in the distribution of the first trigeminal nerve, particularly when there is upper eyelid involvement, the Sturge-Weber syndrome may be associated, with epilepsy and ocular abnormalities. When found in the midline over the lumbosacral spine, there may be an association with abnormalities of the lower spinal cord. When such a lesion feels warmer than surrounding skin and is associated with soft tissue hypertrophy or pulsation, it may in fact be an arteriovenous malformation. Children with these rare lesions should be referred to a paediatrician and they will usually require MRI investigation.

Capillary malformations may be treated by pulsed dye laser, and this is best done as early as possible in the child's life as response is best in the first two years. Such children should be referred to a paediatric dermatology unit at a children's hospital. Treatment usually starts at 6 months of age and, particularly for large lesions, may need to be continued at three to six month intervals over several years. Generally, in 80% of patients the lesion

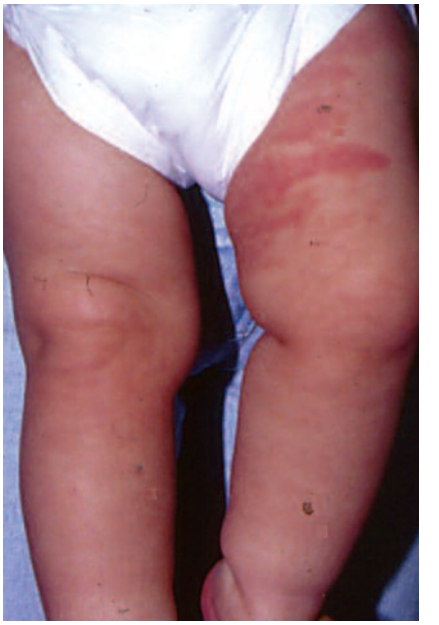


Figure 11. Capillary malformation (port wine stain).

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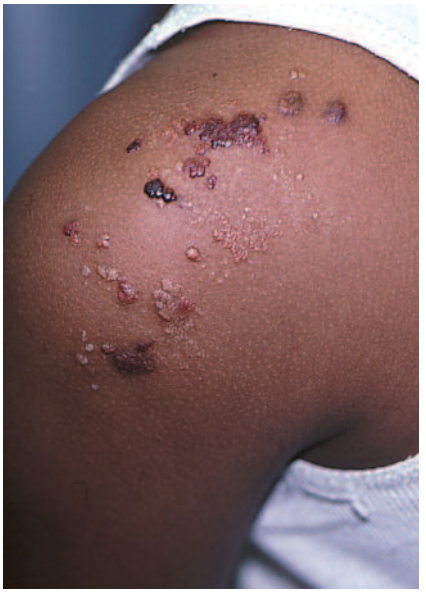

Figure 12. Lymphatic vascular malformation.

will be lightened by 50% or more.

Extensive lesions should be investigated by ultrasound to determine if there are underlying abnormalities. Patients with lesions on the face may require medical imaging and ophthalmology referral.

Other vascular malformations

Other vascular malformations are rare and consist of a mixture of subcutaneous venous and lymphatic elements. Again, they are formed early in fetal life. In the past they were called 'cavernous haemangiomas'. The lesions may be small or large, sometimes infiltrating muscles and joints. The appearance of these is highly variable, but usually there is obvious hypertrophy and sometimes visible enlarged veins, discolouration or bruising. When the lesion is predominantly lymphatic (Figure 12), it may be infected recurrently or leak clear fluid. To determine exactly the nature of the lesion, ultrasound or MRI is required.

Previously, these lesions were generally treated with surgical excision, often with great difficulty and variable results. More recently, interventional radiologists have used sclerosant therapy under fluoroscopic guidance. Some lasers, such as long-pulsed neodymium: yttrium-aluminium-garnet (Nd:YAG) laser, have enough penetrance to treat intracutaneous and superficial malformations. These modalities are available at only a few major centres with specialised expertise.

Conclusion

Although it is unusual for babies to be born with skin lesions, virtually all children will have at least some melanocytic naevi by the age of 2 years. Birthmarks are rarely dangerous and hardly ever have a malignant potential, but they may be cosmetically embarrassing, particularly if they occur on the face. Some birthmarks, particularly the less well-known epidermal naevi, may be mistaken for other lesions such as warts or even dermatitis. Ulcerated haemangiomas may be mistaken for child abuse. Some lesions can interfere with function.

Most birthmarks do not resolve spontaneously in childhood; however, the common haemangioma of infancy always does and so usually does not need treatment.

If they are small, most melanocytic and epidermal birthmarks can be easily removed surgically, with or without the aid of laser therapy, if the child and parents wish it; however, removal of larger lesions can present a significant challenge.

Vascular lesions are often the most difficult to treat because they are too large to be excised and what is seen on the skin surface may be the 'tip of the iceberg'. A combination of surgery, laser and sclero - therapy is used to treat these.

Although malignant potential and interference with function are important, the most common reason for removal of a congenital lesion is cosmesis. With the exception of laser therapy for capillary malformations, which is most effective early in life, the decision to have treatment can, and should, involve the wishes of the child. MT

DECLARATION OF INTEREST: None.