

**Gideon Sandler**

BSc(Med), MBBS(Hons), is Paediatric Surgical Research Fellow, Department of Academic Surgery, The Children's Hospital at Westmead, Sydney, New South Wales. gideonsandler@optusnet.com.au

**Susan Adams**

MBBS(Hons), FRACS(Paed), is Head, Vascular Birth Marks Clinic, Department of Surgery, Sydney Children's Hospital, New South Wales.

**Claire Taylor**

MBChB(Edin), AMC, MS(Trans), is an advanced trainee in urology, Western General Hospital, Footscray, Victoria.

# Paediatric vascular birthmarks

## The psychological impact and the role of the GP

### Background

This study was designed to investigate the psychosocial impact on the family of a child with a vascular birthmark and examine the role of the general practitioner in meeting the family's needs.

### Method

Nineteen families were interviewed with a questionnaire before their assessment at the Sydney Children's Hospital (New South Wales) Vascular Birthmarks Clinic.

### Results

Sixty-eight percent of parents overestimated the size of their child's lesion when asked to draw it; 15% said some lifestyle modifications had to be made to accommodate their child's condition; 63% were concerned about their child being teased at school; and 36% had issues of self blame and embarrassment. Expectations from the clinic included information (68%), treatment (47%) and reassurance (26%).

### Conclusion

Vascular birthmarks are common. Treatment strategies are improving, but there is a need to adequately address the psychosocial impact that these lesions have, both on parents and children. The GP is the carer best placed to meet these needs.

■ **Vascular birthmarks (VBM) are a common heterogeneous group of lesions that affect children from birth or shortly after. They are broadly classified as either haemangiomas or vascular malformations (VMs). Haemangiomas are the most common tumour of childhood affecting 1% of the population. Usually isolated uncomplicated lesions, they grow and regress over time, most disappearing by 9 years of age.<sup>1</sup> Vascular malformations are a diverse group of lesions that have a less optimistic prognosis. Ten times less common than haemangiomas, they grow but usually do not regress, and are more often associated with local or systemic complications.<sup>1</sup>**

The psychosocial impact of children with a VBM on their families is poorly documented in the literature.<sup>2,3</sup> At Sydney Children's Hospital (New South Wales), children with VBMs are managed in a multidisciplinary clinic that offers specialist care: paediatric and plastic surgery, paediatrics, dermatology, radiology, immunology, haematology, pathology, nursing, social work and physiotherapy. Most children are referred from their general practitioner.

### Method

Interviews were conducted with 19 families attending the vascular birthmark clinic (VBC) for the first time. Using a structured questionnaire, the primary or second author interviewed one or both parents before their child was assessed. Notes were taken on the questionnaire during the interviews. Parents were not interviewed in the presence of their child.

Questions pertained to the child's age at diagnosis and at presentation, and the name of the VBM according to the parent. Case notes were inspected to compare the actual diagnosis with the diagnosis as stated by the parents. Occasionally the stated diagnosis changed on the basis of the assessment made at the VBC. Parents were asked draw an estimation of the size of their child's VBM (from memory). Length, width and radial measurements of the lesion and drawn estimation were taken and, according to their shape, their areas were calculated and compared. Parents were asked where they received information about

the VBM, if they had received any conflicting advice, and if they knew anything of the VBM's natural history and treatment. They were asked to comment on what degree family, social and work life had been affected as a result of their child having a VBM. No information was collected with regard to the family's social, demographic or economic situation.

Ethics approval was obtained from the South Eastern Sydney Area Health Service Human Research Ethics Committee (Eastern Section).

## Results

The mean age of the child at first diagnosis was 4.7 months (SD: 16.4); mean age at presentation to the clinic was 25.1 months (SD: 35.5). Most patients had been given a diagnosis before coming to the clinic, but only 15% of parents used the correct term for their child's lesion when describing it. The most common lesion was haemangioma (84%) with two capillary malformations and a nevus flammeus. The head was the most common site of presentation (57%), which included the scalp (21%), lip (15%), eyelid (10%), nostril (5%) and ear (5%) (Table 1). One child had Klippel-Trenaunay syndrome and one had haemangiomas at more than one site.

Before attending the clinic, parents had spoken to and received advice from their paediatrician (84%), friends (63%) and family (47%). Forty-two percent had spoken with their GP.

Only two respondents had done any independent research on their child's lesion before attending the clinic but they knew little about the complications, treatment or natural history of their child's VBM. Overall, only half of respondents knew of the possibility of complications that could arise from the VBM. Fifty-two percent thought laser was a possible treatment option, 42% surgery, and 15% cortisone injection. All but two respondents knew of at least one treatment option that was available to them.

Parents were asked to draw their child's VBM according to its correct size and shape. Sixty-eight percent of parents overestimated its size and 21% estimated the lesion as 3–4 times its actual size. None underestimated the size of the lesion. All respondents approximated the shape accurately. Eighty percent of parents on presentation to the clinic wanted the lesion treated regardless of its site.

Feelings and concerns voiced by parents were varied. The most common issues raised were that their child would be teased at school

and that their self esteem would suffer (63%). Thirty-six percent had issues of self blame and embarrassment as a result of their child having a VBM. This occurred regardless of whom they had spoken to about their child's condition. The most common parental lifestyle modification was taking time off work to attend appointments with health practitioners. Fifteen percent of parents suggested that there had been some modification to family/social life because the lesion was unsightly. One

respondent was afraid that child abuse might be assumed if the lesion was visible.

The questionnaire revealed no notable differences between the parents of children with haemangiomas and those whose children had VBMs, despite the differences in the prognosis of the two conditions. This may be reflective of the small number of participants in this study.

## Discussion

### Haemangioma

Haemangiomas (Figure 1) are benign tumours of capillary endothelium that occur just beneath the skin. They are rarely multiple or affect internal organs. The most common benign tumour of infancy, affecting 1% of the population, haemangiomas are more common in girls; 20% are present at birth and 80% develop in the first 8 weeks of life. There is usually a rapid proliferation phase for the first 6–9 months of life. Growth then slows, and the lesion usually begins to regress after 2 years of age; 90% fully regress by 9 years of age, often leaving a thin, pale area of skin.<sup>1</sup>

Indications for treatment include obstruction of function (eg. occurrence on the eyelid obstructing vision), massive size and occurrence in a conspicuous place (eg. tip of the nose or forehead).<sup>3,4</sup> Current treatment options, if required, include oral or intralesional corticosteroids (60–80% response), subcutaneous interferon- $\alpha$ , vascular specific pulsed dye laser, chemotherapy with vincristine, surgery, or a combination of these. Monitoring should be regular but is case specific. It is required more often in the proliferative phase, if the lesion appears in a danger area (eg. near the airway or eye) and for more anxious parents or carers who need more support. Once stable, monitoring can occur every 6–12 months until resolution.<sup>1,4–6</sup>

### Vascular malformations

Vascular malformations (Figure 2) are a heterogeneous group. Lesions are either comprised of predominantly one vessel type (capillaries, veins, lymphatics or arteries) or a combination of vessels types. Vascular malformations are always present at birth, growth is commensurate with the patient and they do not regress.<sup>1,5</sup> Capillary malformations (port wine stains) are most common, affecting 1 in 1000 people. Peripheral capillary malformations can be associated with hypertrophy of an extremity and 8% of patients with facial lesions have ophthalmologic and central nervous system complications (Sturge-Weber syndrome).<sup>1</sup>

Pulsed dye laser is the mainstay of treatment and the earlier treatment starts the better the response.<sup>1,4</sup> Vascular malformations can be reviewed at the time of treatment. Complex malformations recommend 6–12 monthly reviews for medical as well as psychosocial reasons.

The presence of a VBM psychosocially affects both the patient and their parents. Parents may be subjected to comments, questions and unsolicited advice from friends, family, and complete strangers.<sup>2–4</sup> People may stare, ignore or avoid a child with a VBM. Parents sometimes have to endure suggestions of child abuse. In our study, some parents

Table 1. Location of vascular birthmark

Location	Number of patients (n=19)
Scalp	4
Arm	4
Lip	3
Eyelid	2
Neck	2
Nostril	1
Ear	1
Chest	1
Leg	1

Figure 1. Haemangioma of the leg in a newborn



Figure 2. Capillary malformation (port wine stain) of the face in a newborn



Images reproduced with permission: Auckland District Health Board

modified the times and places that they went out because they felt their child's VBM was unsightly.

School peers can be particularly cruel and insensitive to a classmate with a VBM, which can lead to social withdrawal, anger or aggression, with a significant negative emotional impact.<sup>2-4,7-9</sup> In our study, 63% of parents felt that their child would be teased at school and that their self esteem would suffer. Although children around 1 year of age may be aware of their VBM, they usually refrain from adverse behaviour such as shame or shyness before the age of 4–5 years, particularly if the family have adapted well.<sup>4</sup> However, Weinstein and Chamlin<sup>2</sup> commented, 'unattractive children are more likely to be bullied and teased and to be aggressive and develop antisocial behaviour'. This in part justifies parental fears regarding self esteem and schoolyard cruelty should the VBM not be treated before the commencement of school.

When asked to draw their child's lesion, 68% of the parents overestimated its size with 21% overestimating it by 3–4 times. Within this group were the 36% of parents who identified issues of self blame. Questions such as: 'Did I do something wrong during my pregnancy?' were commonplace during the interviews. Guilt seemed to play a large part in the parental reaction to a child with a VBM, as it if it were somehow their fault.

Having too little information with regards to cause, complications and treatment and not knowing how to respond to the hurtful comments of others may compound parental feelings of uncertainty. The main

sources of reliable information were the GP and paediatrician. But there are often multiple sources of information and, although parents may have been informed as to the name and nature of their child's lesion, they often do not remember the diagnosis.

Communication is paramount and a number of consultations may be required so that parents fully understand their child's condition. Information should be offered in digestible quantities. 'Before and after' photographs may be helpful in reassuring parents and patients about the natural involution of haemangiomas. Contact with parents of other patients may also help such that they may compare experiences and not feel isolated.<sup>2,3</sup> If the GP feels that the family is not coping despite the provision of information and counselling, referral to psychological services may be required.

### Limitations of this study

Parents of patients referred to a specialist clinic may have more concerns as these patients may represent more complicated cases. Relevant findings may have been missed because of the small number of participants.

### Conclusion

Haemangiomas and VMs are common. Despite this, misdiagnoses and misperceptions persist. Further research on this topic is required. Areas for consideration could include perceptions and beliefs of parents before and after attendance at a VBC; perceived accessibility to support services; qualitative assessment of services provided through a VBC; assessment of parental feelings from the time of diagnosis through treatment to resolution (for haemangioma); the value of printed material and, for older children, a separate evaluation of their psychosocial reactions to their VBM, its effect on their family and social life, and how that is modified by the use of intervention.

Finally, GPs and other health providers should be aware of the clinical and social consequences for a child with a VBM. This will enhance the quality of their interaction with the family and lead to timely referral to specialist services and counselling as required.

Conflict of interest: none declared.

### References

1. Morelli JG. Hemangiomas and vascular malformations. *Pediatr Ann* 1996;25:91–6.
2. Weinstein JM, Chamlin SL. Quality of life in vascular anomalies. *Lymphat Res Biol* 2005;3:256–9.
3. Tanner JL, Dechert MP, Frieden IJ. Growing up with a facial hemangioma: parent and child coping and adaptation. *Pediatrics* 1998;101:446–52.
4. Frieden IJ. Which hemangiomas to treat – and how. *Arch Dermatol* 1997;133:1593–5.
5. Enjolras O. Vascular tumours and vascular malformations: are we at the dawn of a better knowledge? *Pediatr Dermatol* 1999;16:238–41.
6. Van de Kerkhof PCM, de Rooij M, Steijlen PM. Spontaneous course of hemangioma: facts and speculations. *Int J Dermatol* 1998;37:101–2.
7. Augustin M, Zschocke I, Wiek K, et al. Psychosocial stress of patients with port wine stains and expectations of dye laser treatment. *Dermatology* 1998;197:353–60.
8. Kassel T. Vascular malformations – how to cope with other people's insensitivity. Information pamphlet, Sydney Children's Hospital's Lymphangioma & Vascular Birthmark Clinic.
9. Kassel T. Vascular malformations – self-esteem and your child. Information pamphlet, Sydney Children's Hospital's Lymphangioma & Vascular Birthmark Clinic.

**AFP** CORRESPONDENCE [afp@racgp.org.au](mailto:afp@racgp.org.au)