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Growing up with Hemophilia

Four Articles on Childhood

by

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GROWING UP WITH HAEMOPHILIA

FOUR ARTICLES ON CHILDHOOD: BABIES

Babies are fun! We know that because Adam, our first grandchild, is busy giving us a refresher course in small people.

When a baby with haemophilia is born he is just as much fun as any other baby. Nothing in his childhood needs to be different.

It is terribly important to realise this. The "START-LINE" for anybody must be normality. This is true of any disorder because if the normal things are not recognised and nurtured they will not thrive. When this happens the disorder can eventually take precedence over everything else, smothering the child and his family and eventually leading to the self recognition of haemophilia as a handicap. A diagnosis of haemophilia is not a handicap; it can only become one if the family or their medical advisers allow it to.

PREGNANCY, BIRTH AND DIAGNOSIS

When there is a family history and the prospective mother knows from this that she may be a carrier, she and her partner should talk to doctors with experience of up-to-date haemophilia care. Any decision about having a child who may have haemophilia always lies with the couple themselves, but doctors can help in the diagnosis of carriership and sometimes, if the couple want it, the diagnosis of haemophilia in the womb. If a twin pregnancy is diagnosed what are the chances of both babies being affected? Identical twin boys will of course both have haemophilia if they have inherited the gene. Non-identical twin boys may or may not both have the condition, it depends on which of the mother's X chromosomes each have inherited.

When a couple decide that they want pregnancy to go to term, arrangements can often be made for diagnosis to be carried out on a specimen of blood taken from the umbilical cord immediately after birth. It is important that good facilities are available both within the maternity hospital for taking the specimens, and within the nearest Haemophilia Centre so that the specimens can be tested very quickly. Any delay may give a false result.

Haemophilic babies are born perfectly normally and are not in any danger during normal vaginal delivery. Caesarian section is only indicated when there are particular difficulties during the delivery, for instance a potentially difficult forceps procedure. On the sixth day of life all babies have blood taken from a heel for the Guthrie test for phenylketonuria. The skin prick is small, and I have never known problems even when the infant has severe haemophilia.

When there is a family history the parents' attitudes to a new born baby with haemophilia will obviously be coloured by the family experiences. If these have been bad with, for instance, particularly nasty bleeds or experiences of surgery going wrong, or premature death of relatives, then the couple will be more fearful for their child than they would be in a family where all the experience is good. Research and development in haemophilia is so rapid that it is very important for couples in this position to get up to date information before their child is born, so they can act on fact and not fantasy. Old medical textbooks from the local library are NOT recommended.

When there is no family history the diagnosis of haemophilia will not be made at birth. It will probably follow either the appearance of bruising when the baby is around one year old and beginning to be more mobile, or perhaps prolonged bleeding following injury in the mouth. Typically, the delicate piece of tissue between the top lip and the gum is injured. In normal children bleeding quickly stops. In a child with severe haemophilia it commonly restarts again because healing is easily disturbed by movement during eating or chattering.

Unfortunately, because haemophilia is so rare (around 1 in 10,000 boys), its diagnosis may be delayed for some time and doctors may look for other causes of bruising including, in some cases, non-accidental injury. This is awful for any parent, but unfortunately reflects our society and the fact that babies who have been injured intentionally form a normal part of the life of any children's department. Clotting tests soon lead to the right diagnosis and referral to the nearest Haemophilia Centre.

Families who have had other children before the son with haemophilia already have experience of the normal growth and development of children, with all the successes and setbacks usual as kids grow up. They can therefore tell the difference between the trials and tribulations of everyday life and the problems caused by haemophilia. The couple with a first born baby with haemophilia have a lot to learn, and life can be especially difficult if they have been blessed with a hyperactive youngster whose only intention is to take command of the family, day and night.

THINGS THAT NEED DOING ONCE YOU ARE HOME WITH YOUR BABY

First and foremost is the need to have a good family doctor and to have access to a good Haemophilia Centre. The family doctor does not need to be expert in the condition but he or she gives all the usual support needed by a young couple and their children. The Haemophilia Centre staff work with the family doctor whilst providing the specialist care for the haemophilia. You should also join the Haemophilia Society.

100,000 genes carry the instructions to bring a baby to life. The only difference between a baby with haemophilia and a baby without haemophilia is that one gene does not work properly in the haemophilic child. I think that the other 99 999 genes deserve priority!

Make sure that you understand your son's diagnosis and are given a "Special Medical Card for Haemorrhagic States", which should be filled in fully. In particular, it is important to know his factor VIII or IX level because on this depends the specific treatment of his disorder. For instance, if he

has a factor VIII level of over 5% he may be able to be treated with desmopressin rather than with a blood product. Throughout his childhood the doctors will routinely test him for inhibitors to factor VIII or IX; the discovery of an inhibitor can mean that a change in treatment is needed.

Like any child, the boy with haemophilia needs to grow up in a secure environment. Injuries are more likely to occur in the cramped, crowded conditions of inner cities with steep stairs and access only to busy streets, than in more spacious houses with gardens. Help can often be given in re-housing by staff of the Haemophilia Centre. Every family with a child with haemophilia should have a telephone in the house, and again Haemophilia Centre staff can often help persuade local authorities to provide telephones for families who could not otherwise afford them.

All parents need to escape the demands of their offspring at times, and the parents of a haemophilic child are no exception. Babysitters can be armed with knowledge of where the couple are, and if they wish the parents can nowadays carry a pager with them whilst they go to the cinema or local pub.

One of the family doctor's roles is to ensure that all children should be immunised against the full range of infectious diseases. When injection is needed, that too is perfectly safe in haemophilia because the volume of the injection is small. My advice is to keep pressure over the injection site for four or five minutes to prevent any untoward bruising. In the case of BCG, for the prevention of tuberculosis, there is no need to put pressure on. In addition to the usual immunisations in childhood all children with haemophilia should be vaccinated against hepatitis B. There is also a new vaccine against hepatitis A and this will soon probably also be recommended to families. No vaccine yet exists for hepatitis C.

Other than the injections needed for immunisation, all other intramuscular injections are banned in haemophilia. Medicines are given by vein instead. Remember that no child should be given aspirin nowadays. This is especially important in haemophilia because the drug interferes with the stickiness of the blood platelets, and adds to problems with bleeding.

It does not matter how babies with haemophilia are fed, although my general advice to all mothers is always to try to breast feed before turning to the bottle. Weaning is normal. Nor is there any reason for handling babies with haemophilia differently than any other baby. Cots do not need to be specially padded and the same care that parents

CHECKLIST

- Enroll with a good family doctor
- Make contact with your nearest Haemophilia Centre
- Join the Haemophilia Society
- Know everything about the diagnosis, including the clotting factor level
- Have a haemophilia card issued for your son
- Learn about the treatment he may need
- Let everybody in the family know about him
- Install a telephone at home and know how to get help when needed
- Go to the baby clinics
- Have him immunized
- Involve his dad, and his brothers and sisters, in his care

take in choosing any toy applies to those chosen for the baby with haemophilia. He should be cuddled and played with, and swung and enjoyed by all the family, relatives and friends just like any other child. There is no need to make any alterations to pushchairs or prams. He can have a baby bouncer and a baby walker, but make sure the walker cannot tip up easily as he becomes more adventurous. If you live in a house with stairs, have a gate fitted at the top of them. In the car make sure that he has a proper baby seat securely fastened in the back. Never travel with him in a front seat.

When I was young clothing for babies was dreadful. Everybody, whether boy or girl, seemed to be attired in white dresses with plenty of lace. This may or may not have something to do with some of the stones we hear about certain sectors of the British establishment! Nowadays clothes for youngsters are super, bright and colourful, warm in winter and cool in summer. That's fine but make sure that, whatever its colour, his footwear is of good quality as he grows up.

SOME COMMON QUESTIONS

Three particular questions that are sometimes raised during infancy concern circumcision, teething, and anal fissures.

Circumcision should not be performed unless there is a real necessity. In medical terms this means good evidence that a tight foreskin obstructs the flow of urine or sometimes becomes inflamed, rather than that the foreskin doesn't retract; that is usual in many boys until they are

around seven years old. If there is a medical reason, or there are irrevocable religious reasons, then the procedure must be carried out very carefully in an operating theatre with full haemophilia protection. This will mean the baby and his mother staying in hospital for a few days.

Babies with even severe haemophilia have no more than the usual trouble when they teethe. Later, when they start to lose their baby teeth all is normal too; the teeth are pushed out very slowly with only a minimal amount of bleeding that does not require treatment.

A lot of babies develop small tears around the anus after passing hard stools. A tear like this is called an 'anal fissure' and it distresses the baby because it is painful during defecation. In haemophilia it can also lead to a little more bleeding than usual. Fissures heal up easily if stools are softened by, for instance, adding more sugar or fruit juice to the diet of a baby being artificially fed. Sometimes a little local anaesthetic cream can help. I have never known a baby with haemophilia and an anal fissure require specific haemophilia treatment.

FOLLOW UP

Other than the need for regular follow-up at the Haemophilia Centre, most families need to spend very little time visiting a hospital in the early years. When they do, it is extremely important that fathers make every effort to get along as well. In our society it is the mothers who are often expected to take their children to hospital for follow-up and to cater for their bleeding episodes. In the old days this attitude used to lead to haemophilic children growing up without real recognition of their dads and with over-dependence on the love and care of their mothers.

*Main lesson of the day: **ENJOY HIM!***

TODDLERS AND PRESCHOOL CHILDREN

Being able to watch young children explore their world provides one of the great rewards of life. Toddlers cheerfully follow a finger - usually into all sorts of potential trouble. Those with haemophilia are no different. They need the stimulus of exploration in order to develop normally. And they need that stimulus in a secure and loving environment. That means a family without fear. Children constantly sense atmosphere and if their parents are afraid of haemophilia, they will be too.

Coming to terms with haemophilia takes time. Occasionally you will need to cry - to mourn the "normal" child you wanted. That's fine. Let it go and then go back to your child, and to the normality of the fun and excitement of watching a new life take shape. You will know you are through the worst when you look at your child and see the smile and not the haemophilia.

Three things can go wrong in those wonderful pre-school years when all parents re-live their early days through the lives of their children. These are accidents, bleeds, and over protection.

ACCIDENTS

Accidents, of course, happen to any child. Most can be avoided. Lock up medicines and household cleaners. Ensure the garden is secure and that dashes to freedom amongst the traffic are impossible. Keep matches out of reach, and use a fire guard. Common sense for any child.

As babies with haemophilia start to become more mobile some parents like to reassure themselves by padding the knees of long trousers, and sometimes sleeves to protect elbows from bruising, but I have yet to see a bleed into a baby's joint as a result of bangs at this age. Bruises are common but are usually superficial and not painful. Whatever you do, don't bandage the joints themselves. That leads to muscle wasting and instability. All babies fall down but they are well padded anyway. Nature provides them with that nice protective fatty layer that we all spend the rest of our lives trying to get rid of.

The only area which is not protected in this way is the head, and one of the worries of growing up with active haemophilic youngsters is head injury. All toddlers bang their heads either by falling or walking into things. However, it is very rare for the ordinary knocks of everyday life to result in bleeding inside the head. I always encourage parents to have a child who has had a BIG bang seen by Centre staff because early treatment will prevent problems. Certainly if a bang to the head has been particularly severe (for instance a hard fall onto concrete, or a child running into the side of a car) he should receive treatment. In these circumstances it is sometimes wise for him to be admitted for a period of observation as well. It must be stressed that injuries leading to this are very rare indeed, and that is one of the reasons I do not recommend helmets for everyday play. The other reason is because the over-use of protective clothing in ordinary activities encourages the development of awareness of handicap.

BLEEDS

Bleeds at this age usually occur as a result of a bang or fall. Superficial bruising is the most common form of bleed. Abrasions and small cuts cause no more trouble than in anyone else, they just need a sticky plaster. Joint bleeds are uncommon in the first three years of life.

CHECKLIST

- Let him play with other children
- Don't be frightened of going out and leaving him with a babysitter
- Let him explore
- Enter him for playschool
- Buy him a tricycle
- Give time to his brothers and sisters
- Involve Dad
- Compare notes with other parents
- Trust your common sense

Admission to hospital for treatment is nowadays extremely rare. If bleeds do require treatment it is given as an out-patient until the family are ready to start home therapy. Problems can arise here because the doctor on call on a particular day may be relatively inexperienced in giving intravenous therapy to small children. Parents should take advice from the staff of their haemophilia centre about the best way of giving an individual child his treatment. In general children should never be taken away from their mother or father; it is far easier to treat a child when he is sitting on his parent's lap.

A small vein set (a fine needle attached to a length of polythene tubing, which is then attached to the syringe) is far and away easier to use than a rigid needle attached directly to a syringe. On no occasion should the same doctor or nurse attempt more than three venepunctures. If he or she fails three times, success is unlikely and a more experienced person should be called. In any case children who have to undergo more than one venepuncture should be rewarded in some way. There is nothing like being out of pocket to a toddler to improve the doctor's venepuncture technique!

Two other points: firstly, parents should know the positions of the best veins to use in their child. Secondly, all this sounds very threatening to new parents, but they can be assured that they will soon get used to the procedures involved and become very skilled at handling their child so that he can have treatment promptly. It is always remarkable to see how children adapt to treatment which, after a while, does not even hurt. It is also remarkable how quickly parents become able to relax and cope with bleeds and with treatment, no matter how scared they are initially of needles.

Pain associated with haemophilia is rare in the pre-school child. When it occurs the best medicine is paracetamol, as in any child. Remember always to keep paracetamol preparations well out of reach; overdose can be lethal. Remember too that medicines containing aspirin are not suitable for children and especially for anyone with haemophilia, because aspirin makes the bleeding disorder worse.

OVER-PROTECTION

TABLE 2	
WHAT'S NORMAL	WHAT'S ABNORMAL
Bikes	Factor VIII or IX Clotting Activity
Clothing	
Going out with other people	
Growth	
Holidays abroad	
Immunisation	
Intelligence	
Jumping	
Kissing	
Life expectancy	
Physical development	
Pig-outs at McDonalds	
Playschool	
Potty training	
Running	
Swimming	
Toys	
Tricycles	

The third thing that can go wrong occurs as a result of fear, especially when parents feel themselves isolated. It is a natural, loving reaction to protect one's children. Overprotection happens when concern about the haemophilia leads to attempts to avoid all the everyday mishaps of normal life. I have come across families too frightened to let their children out of their homes, parents who only let their children play with soft toys and parents who prefer friends not to call because they are so frightened of a childhood scrap resulting in a devastating bleed. The results of this sort of overprotection start to become apparent very early in life and are at their most severe in late adolescence and adulthood. Failure to come to terms with haemophilia leads to isolation, loneliness, and a lessening of opportunities for marriage and a decent career. One of the best ways to avoid overprotection is to encourage activity with family and friends from an early age. Parents should not be afraid of sharing their haemophilic son. All children with haemophilia should be encouraged to participate in activities and sports with others.

SPORTS

The first sport which most children enjoy is swimming. Swimming has just been voted top sport for someone with haemophilia in an international survey of doctors by the World Federation of Hemophilia. Toddlers love going to the pool, and nowadays most neighbourhoods have excellent facilities for parents to take their children to play in the water. Playing in warm water allows exercise of all joints and muscle groups in the body without gravity, and this can be particularly beneficial after a joint bleed. In addition, swimming helps coordination and the development of strong muscles which protect the joints. This in turn lessens the risk of the development of haemophilic arthritis in later life.

No child should grow into adolescence without being able to swim.

ADVICE TO OTHERS

As a child grows he should be allowed out to play with other children and eventually go to playschool. The advice that follows is what I give to all parents.

Firstly, anyone with a responsibility of looking after a child with haemophilia should know that all usual first aid measures apply. Children with haemophilia bleed no faster than other children and there is always time to get help. This is one of the points I always make in letters to teachers. I write these to give to the parents to take to the school themselves. In this way teachers can see that the parents themselves are in control and know all about their son's haemophilia.

When writing to a playschool emphasis should be put on the fact that John can do everything from riding tricycles to bashing other children in the sand pit, and that no special restrictions should be enforced. There are some funny myths about haemophilia, and I think it also worthwhile reassuring teachers that the condition presents no threat to other children. They will be reassured to learn that the great majority of bleeds are internal anyway.

Other than teachers, babysitters and, of course, grannies and other relatives, the only people who really need to know about a child's haemophilia are the family and other doctors who may be consulted, and the family dentist. Going to the dentist should be a regular routine with an older brother or sister or mother or father from an early age. Although nothing will need to be done, the young child learns not to be afraid and has an introduction to dental hygiene which will be of great importance to him in later life. The doctor at your haemophilia centre will prepare letters about your son for his medical and dental colleagues.

Whatever you do, always be open and honest about your child's haemophilia. The more it is talked about the easier life will be. Remember, too that all children like attention and rewards. Both should be distributed evenly throughout a family. Haemophilia should not be an open passport to the sweetie tin! If you find yourself tempted, remember that fat children with haemophilia fare worse than thin children with haemophilia. Extra weight puts more stress on joints, and fat kids are clumsier and more likely to have accidents than thin ones.

Parents who manage to avoid the pitfalls, and look to the normality of their child rather than to his haemophilia, will be amply rewarded.

Main lesson of the day: LET HIM EXPLORE!

GOING TO SCHOOL

Every family album has a first-day-at-big-school picture in it. Tom, aged 4, in school uniform big enough for him to grow into. Beautifully pressed short trousers. Smart school tie. Socks. Clean shoes. It doesn't last long.

First days away from home are always a wrench. Hopefully, by the time of the start of his formal education the boy with haemophilia will have had plenty of opportunity to play and mix with other children. Nursery or playschool should have defused his mother's initial fears of letting other people look after her child - and her child's bleeding disorder. This earlier informal education makes the start of primary schooling easier. Parents will have confidence that their son is able to cope for short periods of time without them. They will know that he can be trusted to the care of others. The boy is used to the discipline of structured play with other children, and will have survived more than one scrap.

Before that first day his parents should have made sure that the teachers are both aware of his haemophilia, and are comfortable with the diagnosis. This can only happen if they have up-to-date information from the family and from the haemophilia centre. All teachers are used to coping with a wide range of disorders at school. For instance, asthma, diabetes and epilepsy are all more common than haemophilia. It really takes an awful lot to faze the average teacher! But they do need that up-to-date briefing, and they need to know what to do if Tom presents them with a problem.

The best way I have found to do this is to give a personal letter about the child to his parents. They then take the letter when they go to see the teachers. The message is clear. They, the parents, are in control of all decisions taken about their son.

A typical letter is shown in the box. It sets the scene for progress by emphasising normality, and stressing the things the boy is able to do with his friends. It starts with some personal details about him. The diagnosis is then given and the fact that usual first aid measures apply is stressed. This is important because all teachers have knowledge of first aid and will be reassured; they don't have to learn anything new.

Some people still think that boys with haemophilia are likely to collapse in pools of blood and spoil the carpet, and the knowledge that most bleeds are internal is important. Because of this the teachers will need to know how to spot bleeds. The knowledge that the boy himself can tell when he is bleeding before there is anything to see will also be reassuring to them. Silly though it seems, some people also think that haemophilia can be caught. Teachers need to be told that it is not contagious and cannot affect other children, if only to reassure their parents. A very important part of the letter is to explain who to contact should any problems arise, and how to do this. I append a card with the names of staff and telephone numbers to the introductory letter. In this way contact can be made directly with the centre if a parent is not available either at home or on a telephone pager. In the UK families with haemophilic children under the age of 16 years can benefit from a valuable free service called "Armourpage", which is sponsored by the Armour Pharmaceutical Company in association with B.T. and the Haemophilia Society.

PRIMARY SCHOOL	
<i>He Can</i>	<i>He Shouldn't</i>
Play in the playground	Box
Do physical education	Play rugby football
Swim	
Run	
Cycle	
Play football	
Go on school trips	
Do metalwork	
Do woodwork	

The next section of the letter deals with activities and sports and again stresses normality. Normal development depends on continuing exploration of the world. Only the individual can eventually decide what is right for him in terms of work and leisure. Unnecessary restrictions hinder this progress. Children wanting to do the same things as their friends will eventually either reject restrictions directly, or hide the fact they are disobeying their parents in order to avoid retribution. Far better to let them learn within sensible limits what is right for them and for their haemophilia. The only restrictions I recommend are that boys should not box or play rugby football, and the letter explains why. Finally, I ask to be kept up to date with the boy's progress. This is because I want to make sure that decisions which could affect his future and which are unduly influenced by the diagnosis are not taken at school.

In addition to the letter and the card, Haemophilia Society literature about schooling is helpful, as are some of the leaflets now available from the pharmaceutical companies. Occasionally either the parents or the teachers ask for all this information to be followed up by a visit to the school by a member of the centre staff. This can be extremely helpful if there are special difficulties, for instance the presence of high titre inhibitors.

Nowadays the great majority of boys with haemophilia can attend normal school and compete as equals with their peers. It is not very long ago that this was not the case, and in some countries it is still more usual for special schooling to be recommended for haemophilia. If this happens it is very important for the family to discuss the recommendation with centre staff and with representatives of the Education Authority before making a final decision.

The boy with haemophilia deserves the best education that is available. His future happiness and prosperity depend on schooling which has allowed him to thrive outside the diagnosis of haemophilia, which is only one of the many facets that influence decisions about an eventual career. The choice of that career is crucial to him. Although modern treatment allows him to do most jobs, whether they depend on strength of mind or strength of body, he will be competing with others without haemophilia. He has to prove to himself and to others that he is the best candidate for the job, with or without the bleeding disorder.

*Message of the day... **plan ahead for his future.***

CHECKLIST*What teachers need to know:*

- His personal details
- The diagnosis
- The severity
- First aid recommendations
- Who to contact and how
- What treatment is given
- Will special provision be needed in school
- Any effects on other children

I thought I should drop you a note about Tom. He is a fine boy who is developing normally. He has a younger sister, Anne, and the family live in Newtown, where his father is a plumber. You will know from his parents that Tom has severe haemophilia. This is caused by the deficiency of one of the blood clotting ingredients. Because of this deficiency blood does not clot as quickly as it should following injury. Children with haemophilia bleed no faster than other children; their bleeds simply last longer. This distinction is an important one. It means that there is always plenty of time to seek help if it is needed, and it means that the first aid measures used with any child who is injured still apply. Most bleeds in haemophilia are internal.

If they show at all it is usually as bruises, most of which are superficial and of no consequence. Bleeds into muscles and joints do need treatment. Tom knows when he is bleeding and will tell his teacher. Treatment of haemophilia itself is easy. The missing ingredient is replaced in the form of a blood product given into a vein. Tom's parents have been taught to do this themselves. If they are not available when Tom needs them, all you have to do is to ring the Centre; the names and telephone numbers of staff are appended. A member of staff is always available during school hours. Tom is a healthy and active little boy. His parents have been very sensible, and have not tried to restrict his activities. We advise that he should be allowed to participate in everything his friends do at school. As he grows, he should be encouraged to develop skills in sport (especially swimming and team sports) and physical education, as well as crafts like woodwork and metalwork. If trips away from school are planned, the address of the nearest haemophilia centre and the name of the doctor concerned are available. For long trips or visits to other countries, we will supply a holiday kit, together with a letter for the Customs, details of treatment facilities, and, when required, help with travel insurance. The only restrictions affecting Tom are that he should not box or play rugby football. This is because of the special risks to the head and neck involved in these sports. Head injury in haemophilia can be serious, and immediate referral to the centre is necessary.

ADOLESCENCE

It is a tribute to human resilience that most youngsters and their families survive adolescence. Being a teenager is difficult enough without the burden of haemophilia. Whilst parents need patience, a sense of humour and a bottomless bank balance, their son needs both the security of home and the space to grow to the independence of adult life.

There is one word which sums up everything families needs to know about adolescence and haemophilia. It is CONTROL. When someone is in control of their health life is normal. When health is compromised and in control of them, life is abnormal. If someone with haemophilia allows his disorder to govern his life he is not in control; his haemophilia is, and his happiness is clouded. Learning to put life first and haemophilia second is the single most important feature of growing up.

To be in control requires discipline. It is useless to simply "forget" haemophilia and not to, for instance, have a dose of factor VIII or IX before an event known to provoke a bleed. It is silly to imagine that a run of bleeds into a major joint will just "go away" without therapy or subsequent damage. It is foolhardy to go off on holiday without packing the equipment needed to treat a bleed, or finding out the location of the nearest haemophilia centre. In all these cases common sense planning ensures that life can run smoothly without the disruption of untreated haemorrhage.

Most children will have learnt to control their haemophilia before puberty. They are in command, knowing exactly when to ask for treatment, judging the dose, mixing their concentrate, increasingly doing their own venepunctures, and clearing up afterwards. And then forgetting about it and getting on with the far more exciting events of life with their friends at home and school.

If this hasn't happened haemophilia can make adolescence very difficult for a while. At some stage there has to be a break. No-one wants haemophilia, but it is there. Coming to terms with it whilst at the same time wanting to slip the net of parental influence, can be very frightening and hurtful. The obvious response is anger, and anger used to be a frequent occurrence in the consulting room before comprehensive care became established and families learnt about letting children taking responsibility for their haemophilia gradually as they grew up.

CAREERS

Choice of career is especially important in these days of high unemployment. Few jobs are closed to those with severe haemophilia; they include work involving particular hazard (for instance the armed forces and emergency services) or the risk of being isolated from special medical help should an emergency arise. With these exceptions and contrary to popular opinion, people with severe haemophilia are able to work manually, and some of them undertake the sort of work that would rapidly floor most doctors. Obviously, whatever career is chosen the man with haemophilia must be ready to compete as an equal to somebody without haemophilia, both when applying for jobs and when working.

There are obvious benefits in becoming as highly qualified as possible before leaving school or university; the choice is so much wider. All schools have easy access to advice about careers, but sometimes this is given rather late in the day. In the context of severe haemophilia it is

important that the possibilities are explored sooner rather than later. Early planning helps ensure that any disruption caused by untoward bleeding is catered for, and special help with difficult subjects is provided. Most youngsters need time to develop their own ideas about what they want to do with their lives; early decisions can always be changed later in the light of new interests and work experience. When doubt about choice of career persists a consultation with an educational psychologist may help to point an indecisive teenager in the right direction.

SEXUALITY

The knowledge that haemophilia is inherited and that the abnormal gene can be passed on to children is bound to colour attitudes to sex and parenthood. Young men who have not yet come to terms with their haemophilia feel more "different" than is usual in adolescence, and may find early courtship difficult because of this. Carrier girls are vulnerable because of the possibility of having haemophilic sons. Life is especially hard for them if there is a bad family history, or if Dad's haemophilia is complicated by severe arthritis, hepatitis or HIV infection.

Two aspects of adolescence may need sensitive counseling. Firstly, carrier girls may have low factor VIII or IX levels and these may be linked to heavier periods than usual. When this happens it is very easy to put things right - one of the low dose pills, or regular courses of cyklokapron, are all that is needed. Secondly, young men may have bleeds induced in the heat and passion of the moment. Again, treatment is straightforward, painful bruising being countered by a dose or two of the relevant factor concentrate. One event that is fairly common at this age is a psoas bleed - that needs energetic treatment with replacement therapy for several days, together with bed rest and then physiotherapy. The psoas muscle runs from the side of the backbone and sweeps forwards round the pelvis to emerge in the front of the thigh. If not stopped quickly bleeding into it compresses the major nerve to the quadriceps group of muscles leading to weakness, instability of the knee and a loss of sensation to touch which can extend from groin to foot. Recovery from a poorly treated psoas bleed can take many months.

SPORTS

Keep active : keep well! Physical activity promotes powerful muscles which protect the major joints. Flabby muscles cannot support joints properly and bleeding episodes are more frequent. Obesity adds to the problem by putting more strain on the unstable joints.

Children of all ages should be encouraged to participate in sports. By the time the boy with haemophilia has reached adolescence he should know which activities he enjoys. He should also be learning those activities which result in bleeds and, if these occur during a sport he has set his heart on, whether he can control them with prophylactic factor VIII or IX beforehand. Regular, enjoyable sport results in feelings of well-being and fulfillment, which help counterbalance the unease and loneliness that haemophilia can provoke. Team work is another powerful weapon in dispelling ideas of isolation or handicap.

Recently, on behalf of the World Federation of Hemophilia, I asked colleagues for their views on sports most suited to people with severe haemophilia A or B. All agreed that swimming, table tennis and walking were to be encouraged, and many more sports were recommended. The "top 10" sports are listed in the first table.

In the second table are the least recommended sports, some of which carry a major threat of head or neck injury.

Altogether the questionnaire listed 69 sports. Bungee jumping was not one of them, although I know three people with haemophilia who have tried it. It would terrify me, and is probably the quickest way of finding out if there are weak blood vessels in the brain - prophylaxis is advisable first! Football (soccer) was well down the list; some doctors thinking that at a competitive level the risks of damage to the legs is too high. I do not share this view, preferring to let youngsters make up their own minds. What is essential for football, as for any sport, is that the usual protective clothing is always worn. No self respecting batsman (if there are any left) would dream of facing Merv Hughes without all the gear.

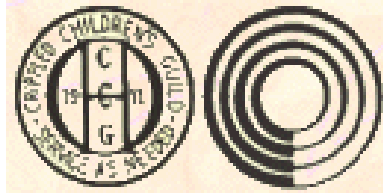
TRAVEL

During the teens most youngsters make their first escape from parents and siblings. Holidays abroad, summer camps and trips with the school all provide the necessary inducements. Haemophilia is not a bar to travel. Again, common sense precautions ensure the trip is a happy one. A holiday letter detailing the medical history and the usual haemophilia treatment should be obtained from the centre which should also provide a clearance note for customs for any equipment, concentrate or desmopressin (DDAVP) carried. A list of centres nearest to the destination, holiday insurance (read the small print!) and, in Europe, Form E111 are the other essentials. And don't forget that lads (and lasses) with the haemophilia gene are just as likely to fall foul of alcohol, drugs and sexually transmitted diseases whilst in Benidorm as anyone else.

Finally, having survived parents, brothers, sisters, schoolmasters doctors, the dreaded nurse, **AND** adolescence, the average young man with haemophilia wants nothing more than to live his life his way. Great! But one thing to think about please. Others less fortunate than yourself (especially those with haemophilia in developing countries) now need your enthusiasm, your energy and your expertise in living with haemophilia. The various haemophilia societies and the World Federation of Hemophilia will welcome your active participation. So add them to your list. Greenpeace, Save the Whales, Antivivisection... and the Haemophilia Society.

Main lesson for life : maintain your CONTROL.

The top 10 sports recommended by doctors. All 100% of those replying put swimming at the top of this list; 94% recommended cycling.	The least favoured sports. All the doctors were opposed to boxing and 90% thought skateboarding dangerous for someone with severe haemophilia.																																												
<p>TABLE 1</p> <table border="1"> <thead> <tr> <th>SPORT</th> <th>%</th> </tr> </thead> <tbody> <tr><td>Swimming</td><td>100</td></tr> <tr><td>Table Tennis</td><td>100</td></tr> <tr><td>Walking</td><td>100</td></tr> <tr><td>Fishing</td><td>99</td></tr> <tr><td>Dance</td><td>98</td></tr> <tr><td>Badminton</td><td>98</td></tr> <tr><td>Sailing</td><td>98</td></tr> <tr><td>Golf</td><td>96</td></tr> <tr><td>Bowls</td><td>95</td></tr> <tr><td>Cycling</td><td>94</td></tr> </tbody> </table>	SPORT	%	Swimming	100	Table Tennis	100	Walking	100	Fishing	99	Dance	98	Badminton	98	Sailing	98	Golf	96	Bowls	95	Cycling	94	<p>TABLE 2</p> <table border="1"> <thead> <tr> <th>SPORT</th> <th>%</th> </tr> </thead> <tbody> <tr><td>Boxing</td><td>100</td></tr> <tr><td>Rugby football</td><td>99</td></tr> <tr><td>American football</td><td>99</td></tr> <tr><td>Karate</td><td>94</td></tr> <tr><td>Wrestling</td><td>93</td></tr> <tr><td>Motorcycling</td><td>91</td></tr> <tr><td>Judo</td><td>91</td></tr> <tr><td>Hang-gliding</td><td>90</td></tr> <tr><td>Hockey</td><td>90</td></tr> <tr><td>Skateboarding</td><td>90</td></tr> </tbody> </table>	SPORT	%	Boxing	100	Rugby football	99	American football	99	Karate	94	Wrestling	93	Motorcycling	91	Judo	91	Hang-gliding	90	Hockey	90	Skateboarding	90
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The Hemophilia Treatment Center, Orthopaedic Hospital,
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of the World Federation of Hemophilia.
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