



Intrauterine Growth Retardation (IUGR) including Russell Silver Syndrome

A Guide for Parents and Patients



SERIES 1



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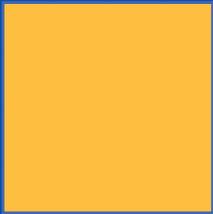
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This booklet is intended to provide help when dealing with problems or difficulties associated with your child's condition and to provide information which will enable you to understand your child's treatment better and give you a basis for discussions with your child's specialist when necessary.

If you require further general information about *Intrauterine Growth Retardation*, you can contact the Child Growth Foundation.



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INTRAUTERINE GROWTH RETARDATION – Series No 14 (January 1997)

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Thanks to Dr David Dunger (John Radcliffe Hospital, Oxford) for his kind assistance in editing this book.

The British Society for Paediatric Endocrinology and Diabetes (BSPED) is an association of specialists who deal with hormone disorders in children.

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INTRAUTERINE GROWTH RETARDATION

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INTRAUTERINE GROWTH RETARDATION

A low birthweight, or small for dates, baby is defined as a baby born with a weight that is inappropriately low for the duration of the pregnancy – for a baby born at term this would be a birthweight less than 2.5 kg. The inappropriately low weight indicates that the growth of the baby in the womb has been unsatisfactory and this is why it is called Intrauterine Growth Retardation, abbreviated to IUGR.

The majority of babies born small for their gestational age show catch-up growth over the first two or three years of life. However, in about a third complete catch-up growth does not occur. These children remain small and fail to reach their genetic potential as defined by their parental heights.

This booklet is concerned with babies born small for their gestational age due to intrauterine growth retardation including Russell Silver syndrome. We have intentionally left out information on babies born small for gestational age with other rare syndromes as in such cases the problems are often more complex. Parents wanting information about these conditions should get in touch with the Child Growth Foundation and be directed to the appropriate parents' support group.

Low birthweight babies may be thin and under weight for their age and may experience some of the problems associated with Russell Silver syndrome but often lack the distinct physical characteristics which are linked with this diagnosis. These babies need to be monitored by a paediatrician and referred to a growth specialist if their growth does not progress towards normal. The option of treatment to help with their growth may be discussed. Though your child may be diagnosed as having intrauterine growth retardation, you may recognise some of the features described in the section on Russell Silver syndrome as some of the problems do overlap.

RUSSELL SILVER SYNDROME

This condition was first described by Dr Russell in England and Dr Silver in the USA in 1953/54. At first it was thought that they were describing different syndromes but then it became clear that they had seen aspects of the same condition. The syndrome is called Russell Silver in the UK and Silver Russell in the USA! Russell Silver syndrome is very rare, occurring in 1/50,000 to 1/100,000 births. However, it is probable that there are many conditions which are similar and which we describe collectively as Russell Silver syndrome and this may lead people to think it is more common than it is.

Little is known about the cause of this condition and why some children with IUGR have specific features of the Russell Silver syndrome and others do not. In the majority of families only one child is affected but very occasionally families do have more than

one affected child. This may suggest a genetic basis for this condition and this is currently the subject of much research.

In some children with the milder forms of IUGR (but not Russell Silver syndrome) a genetic irregularity has been found. These children have two copies of one chromosome (chromosome 1) from their mother instead of one copy from each parent. However, the significance of this finding is not known.

It is recommended that families who have a child with Russell Silver syndrome seek the help of genetic counselling before they decide to have any more children.

DIAGNOSIS

The diagnosis of intrauterine growth retardation is based on careful comparison of the baby's weight centile at birth with standards which take into account the gestational age of the baby (i.e. the length of the pregnancy). Ideally, infant length should also be measured but this needs to be done accurately, using the correct equipment, if the measurement is to be meaningful.

The diagnosis of Russell Silver syndrome is based on these same measurements with the following additional observations. (NB: At present there is no special test which confirms this diagnosis and so it is made on the basis of physical characteristics.)

The main points to be established from the medical history are:

- IUGR can often be identified in early pregnancy
- Early feeding problems are common. The baby is often disinterested in feeding and takes only small amounts with difficulty.
- These babies often sweat a lot, particularly at night, and have a greyness, or pallor, of the skin. In some infants, this is a symptom of a low level of sugar in the blood (hypoglycaemia). As they get older, these infants/children may have altered behaviour, such as hyperactivity or, conversely, tiredness, as a symptom of their low blood sugar level.

Physical features are often not clear until after the first year of life. On examination, some of the following may be seen (**NB: it is important to remember that your child is very unlikely to have all the features described**):

- A small triangular shaped face with a small jaw and a pointed chin.
- The small jaw may subsequently lead to dental problems due to crowding of the teeth.
- A mouth which tends to curve down.
- A blue tinge to the whites of the eyes in younger children.

- The head circumference may be of normal size, which means it can appear large in comparison to the small body size. This may result in the doctor checking for hydrocephalus (a build-up of fluid surrounding the brain) although this problem is not associated with the diagnosis of IUGR or Russell Silver syndrome.
- The opening between the bones of the skull, the fontanelle, may be very wide and late to close. (NB: This opening is under the skin at the top of the skull and is there in all babies.)
- The little finger of each hand may be short and curve inwards (clinodactyly). This is the most frequent distinguishing feature of Russell Silver syndrome.
- Body asymmetry is where one side of the body grows more slowly than the other. The result is that one side of the body, sometimes including the face, is smaller than the other.
- Continued poor growth with no 'catch-up' into the normal centile lines on a height chart (your doctor can show you this).
- Puberty may commence at an earlier age than normally expected.

Often it is not until after the first year of life that the characteristic features of Russell Silver syndrome are recognised as being present.

GROWTH

In this section, the term IUGR is used to cover both the low birthweight babies and those with Russell Silver syndrome, as there are many features that they share.

Most of the children with Russell Silver syndrome, and about one-third of those with severe intrauterine growth retardation, fail to show catch-up growth by two to three years of age. The remainder of those with IUGR tend to have caught up to their expected height centile within two to three years.

Growth is mainly dependent on nutrition during the first year of life and one of the characteristics of children with low birth weight is that their intake of calories may be insufficient as a result of their feeding difficulties (see later section). After the first year of life, the role of growth hormone becomes more important in growth.

The likelihood of good catch-up growth is much greater if the intrauterine growth retardation develops during the later stages of pregnancy. In contrast, IUGR which occurs in the first or second stages of pregnancy often results in a limited or absent catch-up growth. This failure to catch up is not helped by the feeding difficulties, and consequent reduced calorie intake, which are seen during the first year of life.

In those children who do not show catch-up growth during the early part of childhood, short stature remains a problem. Puberty tends to start around the normal time, or very slightly early. The pubertal growth spurt may be less than anticipated and so final adult height may not be as good as would have been expected, even for their low birthweight. Young children with Russell Silver syndrome often have a delayed 'bone age' which is often used as an indicator of their physical maturity.

However, in some IUGR children, and for reasons we do not understand, the bone age advances inappropriately during the middle childhood years and the bones stop growing at an earlier than expected age. In such cases the child may not achieve their predicted final height.

The reason why growth remains abnormal in children with IUGR is not well understood, although growth hormone deficiency/insufficiency can occur in this group of children. There are various reports as to how common this may be and the range seems to be from 10 to 30% depending on who has conducted the study. So, as part of the investigations of their poor growth, these children will usually have a test of their ability to produce and secrete growth hormone. Because these children often have a low level of sugar in their blood normally, extra care should be taken if they are having a test of growth hormone secretion.

FEEDING

An additional problem in the early months of life is that these children tend to remain very thin, often associated with the feeding difficulties, and so they do not build up fat reserves. This means that they continue to have a lower than normal level of sugar in their blood, and so are at risk of having hypoglycaemia, particularly when they are ill. With some IUGR babies, the specialist will advise the families to try intensive feeding,



A child with Russell Silver syndrome

through a nasogastric tube, to ensure that the baby is receiving enough calories. Although effective, this naturally can be distressing for both the infant and the parents.

Babies who are born with severe IUGR often find it difficult to breast feed as they do not suck easily and they tire quickly. Thus, bottle feeding may be the more successful option for mother and baby. However, many mothers have persevered and have successfully breast-fed their child under difficult circumstances. You will need to decide which is easiest for you and your baby.

Although these babies may eat an amount of food suitable for their size, it is worth trying to encourage them to increase slightly the intake of calories.

However, if forced to have too much, even by well-meaning professionals who are trying to encourage weight gain, these babies often gag/vomit it all up which not only distresses you, but may negate the benefit of the whole feed.

Babies with IUGR often have a high arched palate (roof of the mouth) and so, for these infants, sucking and eating may be difficult. This can mean that when they are supposed to have moved to solid foods they may find it difficult and spit out even the tiniest lumps or solids. It may therefore be easier to continue with pureed food until the child is a little older.

In addition to feeding difficulties, your child may have constipation or, conversely, diarrhoea. Either of these problems will need to be treated before anything can be done successfully to address the feeding problems. Speak to your GP and your specialist to make sure this isn't a problem, or that it is treated properly.

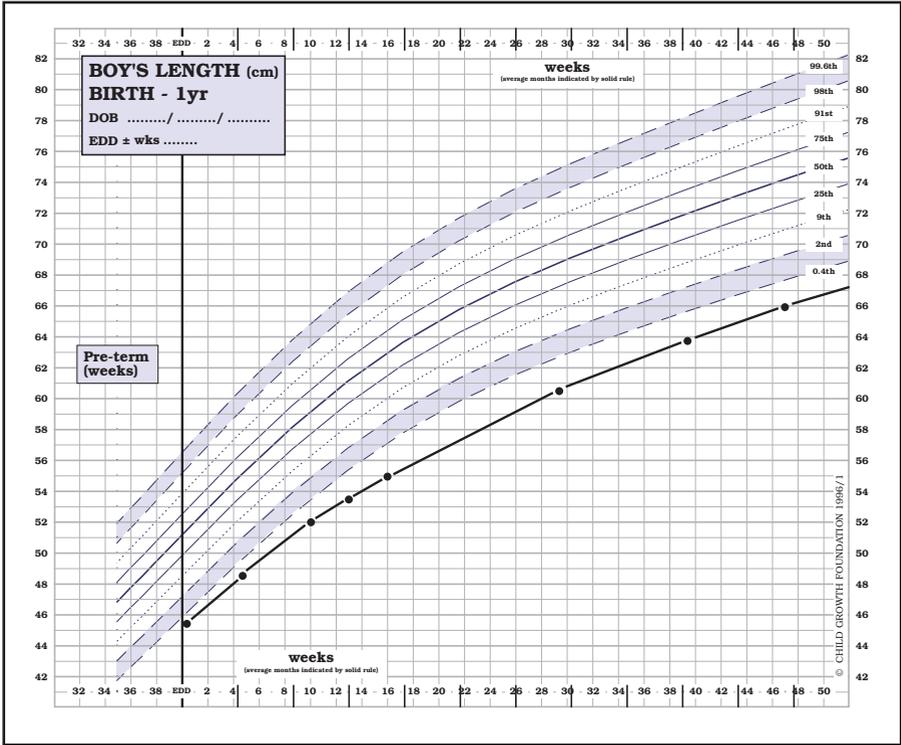
Introducing solids

Even when very young, these children can be very disinterested in feeding – and they can show a real determination not to eat solid foods. This may, in part, be because they associate this with the unpleasantness of gagging, even on the tiniest lumps. Also, it may be because eating solid food requires different mouth and tongue movements to eating liquids or purees.

However, it is important that your child learns how to eat solids and it may help to start with ones which can be dissolved in the mouth, e.g.:

- Small pieces of chocolate
- Weetabix
- Savoury snacks, such as Quavers

Naturally, parents are very concerned that their child has the right intake of calories and so the temptation may be to concentrate only on sweet foods. However, it is also



A typical growth and weight chart for a boy with Russell Silver syndrome

important to achieve a balance to your child's diet and so savoury foods do need to be included. In fact, some children with IUGR/Russell Silver syndrome seem to prefer salty or spicy foods. Other ideas to increase the calorie value of food for your child include adding full fat Jersey milk, or even cream, to drinks, breakfast cereal, custard, etc.

Once your child has started to take solid food, and this may be slightly later than with other children, you may notice that their diet is not very varied. This is common with IUGR children as they often stick to their preferred food and so can seem to be very fussy in their eating habits. This may improve as the child gets older, but eating little for a few days and then appearing to have an insatiable appetite may continue until after the child has gone through puberty. You will soon discover what your child's favourite foods are. As mentioned above, some of them may be spicy and/or salty: this seems to be quite common.

Ask for help and support from your health visitor and GP as they may have ideas to help you overcome some of the feeding difficulties. This will be helped further if your specialist gives your GP as much information about the condition as possible, particularly whether your child has a high arched palate. Alternatively, a speech therapist with experience in feeding difficulties may be very helpful. Ask your GP or specialist if there is one in your area.

Maintaining food intake and avoiding hypoglycaemia

Children with IUGR who remain small generally have small appetites and eat small quantities at one meal but may want to eat more often, compared with children of the same age. These feeding difficulties, with low food intake, can lead to low levels of sugar in the blood. This can cause excessive sweating, particularly around the head and upper body, which can be quite alarming for the parents and distressing for the child. Your child may also become very tired and irritable if the level of sugar in their blood is low. This problem will usually improve as your child gets older but can reappear if your child is ill.

If there are regular or more severe problems, admission to hospital may be needed to check the level of sugar in the blood and this may require an overnight stay and increased calories before bedtime (e.g. chocolate biscuits or bananas) may be recommended.

It may, therefore, be important that your child has access to biscuits or snacks at school. If it is not your school's policy to allow this, your specialist or dietician will be able to help by advising the school of the necessity. After all, it is in the school's interest too that your child is happy and alert during classes!

Your child will be thin and underweight for his/her age, and the lack of subcutaneous fat may make you feel that your child needs to eat more. Thus, when little interest or 'faddiness' is the only reaction to food that your child shows, meal times can become a

time of confrontation. It is important that this is avoided if at all possible and, so as long as your child is alert and active, the smallness of their appetite will not cause any harm. If a low level of blood sugar is causing irritability, a biscuit or a fruit drink will help. It is more important that you encourage your child to eat small amounts of food regularly than a large amount at any one particular meal.

SCHOOLING

Most children who are born with intrauterine growth retardation are of normal intelligence and go through mainstream schooling. For those children who do have special educational needs, this mainly relates to difficulties with concentration, organisation and problem solving. If you feel that your child has a learning difficulty, you should discuss this with their teacher to see whether an assessment from an educational psychologist should be sought. Your growth specialist may be able to help support your request through a letter indicating the nature of the condition and why there may be special educational needs. Children who need extra help could benefit from 'statementing' of educational needs and the identification of extra support within the normal school system.

Young children who are small for their age may have difficulties coping in surroundings which are designed for children who are taller and bigger. The height of chairs and desks need to be at a height that will allow them to see the board and to write easily. Also, they will want to be able to reach coat hooks and toilet locks without always asking for help. It is worth discussing these issues with the form or head teacher before they become a problem for your child.

Clearly, a small child may resent being treated as younger than his/her years and as being less responsible than he/she is. However, it may be less clear that a child who is always treated as being younger than their years can fall back and behave according to the age that their size indicates and not their true age. This can apply both educationally and emotionally.

Such a child may therefore achieve much less than their true potential and so lose self-confidence in their abilities. School is as much about building confidence and self-esteem as it is about dependence and trusting relationships. Developing these abilities can seem impossible to the child who is constantly having expectations of his/her abilities lowered.

It is very important that teachers are aware that the child needs to be treated according to their age and not to how old they appear to be, while not expecting them to undertake activities that are inappropriate for their size. In addition, parents too need to be sure that they are not treating their child inappropriately for their true age.

PHYSICAL EDUCATION/SPORT/GAMES

Sports and games are generally perfectly safe for a small child but there may be certain activities which are much more difficult for them and they should not be forced to participate. With the co-operation of your school you will, no doubt, identify those activities where your child can take part and do well, and these may include swimming, gymnastics, etc., rather than contact sports such as hockey, football or rugby, at least until they are older.

OVER-PROTECTION

Even though your child is small, they are still able to take on responsibilities for themselves and others and this, as well as an appropriate level of independence, should be supported. These skills are acquired over a period of time, they do not happen overnight! Your child will need your encouragement to achieve this; if someone is always doing things for them they may find it more difficult to accept appropriate challenges as they get older.

Try to ensure that your child is treated according to his/her age, not size. As with any child, they need understanding and encouragement to achieve their potential.

BULLYING

Although children may experience bullying at school, there are some who seem to be more vulnerable than others. Any child who says they are being bullied should be listened to very carefully. Talk to you child's form or head teacher as every school should have a policy for dealing with bullying.

OCCASIONAL PHYSICAL FEATURES SEEN IN CHILDREN WITH SEVERE IUGR/RUSSELL SILVER SYNDROME

Some of the features which will now be described may have only minor medical significance but can be of major importance to the individual concerned. We have tried to give explanations and practical advice where possible. **It is unlikely that any child is going to have all the features mentioned, only a combination that is personal to them as an individual:**

Hypospadias

Very rarely, in boys, the opening of the urethra – the tube leading from the bladder to the opening in the penis – may be short and so not reach to the end of the penis as it

should. This can be overcome by surgery to extend the urethra to its required length. You may wish to ask your specialist how old your child needs to be to have this operation. It is important that your son is not circumcised as the foreskin may be needed for the surgical repair.

Undescended testes

Some boys have this problem whereby the testes have not descended into the scrotal sacs. This may be treated with medication or, if that is not appropriate, with surgery. This will usually be after the boy is two years of age.

Bowel problems

Although bowel problems are not recognised as being part of the Russell Silver syndrome, many parents have commented that their children have experienced constipation as toddlers which can be difficult to cope with. Other children, however, have diarrhoea but this seems to improve as they get older. Your GP or specialist may be able to offer advice as to any treatments which could help.

Hyperactivity

From their early years, these children are extremely active and may find it hard to concentrate. **When they go to bed they often are unable to calm down and have difficulty sleeping.** If your child is particularly hyperactive, and you need a break, it may be possible to arrange for respite care. This should be discussed with your specialist, your GP and your Health Visitor. There are also organisations, such as Home Start, which help families who have any children under school age and arrange for trained volunteers to come to the home and help according to the needs of the individual family.

Delayed development

Babies with IUGR/Russell Silver syndrome sometimes have difficulty lifting their head and show a lack of balance. In infants and children, the development of motor skills, such as co-ordination, may be slightly delayed, because of their smaller size, and so they have reduced muscle strength compared to children of the same age. This needs particular awareness on the part of teachers so that a child is not put in a situation where they can't physically cope. This does not mean that they should be over-protected: rather that they are helped to develop skills but not forced into activities that are outside their physical capabilities.

Hypoglycaemia

Hypoglycaemia is another name for a low level of sugar in the blood. The symptoms of this can be irritability or sleepiness. Many children with IUGR can become very irritable after physical activity, such as games or swimming, at the end of the school day, and/or on waking up. Because these children are thin, and do not have much subcutaneous fat, the

level of sugar in the blood can fall very quickly and this causes the irritability. Giving your child a biscuit or a sugary drink will raise the level of sugar and they should become better tempered within five to ten minutes. With a baby, dipping your finger in honey or glucose and rubbing it round the inside of their mouth will have the same effect.

If your child becomes sweaty, grey and/or sleepy when you would expect them to be awake and possibly hungry, it may be because of a low level of sugar in the blood (hypoglycaemia). It is important that you wake your child and give him/her something sugary to eat. If your child has these periods of inappropriate irritability/sleepiness, it is important to let your specialist know.

If low levels of blood sugar continue to be a problem, some children may need to be given emergency hydrocortisone treatment, particularly during stress or illness and especially if they are vomiting. You will, therefore, need to be aware of how to give emergency hydrocortisone treatment (Booklet No. 5: *Emergency Information Pack for Children with Cortisol and GH Deficiencies and those Experiencing Recurrent Hypoglycaemia*). **It is also important that teachers are aware of the signs of hypoglycaemia in your child.**

Emergency treatment for hypoglycaemia using injections of glucagon should not be used in children with Russell Silver syndrome. This is because glucagon works by releasing stores of glucose from the liver into the bloodstream but, in Russell Silver syndrome, the stores of glucose in the liver are usually already used up. Only hydrocortisone should be given.

It may be that your child additionally has growth hormone insufficiency which will need to be assessed and which may also increase the likelihood of hypoglycaemia.

Protruding ears

This may be a problem and if of particular concern, plastic surgery is an option.

Hearing and speech

Ear infections are quite common and, if persistent, may cause loss of hearing which in turn could affect speech development. If you suspect any loss of hearing in your child it should be quickly investigated and referral made to an ear, nose and throat specialist. Help from a speech therapist may be very beneficial if there is any difficulty with talking.

Body asymmetry (Russell Silver syndrome only)

Sometimes the difference in length between the right and left legs is so small that it can be managed easily by inserting a thick inner sole into the appropriate shoe. If the difference is more marked, a referral to an orthopaedic surgeon should be made. Your child's growth will be monitored and surgical options such as limb lengthening will be

considered. NB: There are some orthopaedic surgeons who will suggest that the longer leg should be shortened; this is not the appropriate approach and so your specialist will help you with the appropriate advice.

Your child's feet may be of different sizes and there are some firms that try to help with this situation so that you don't have to buy two pairs of shoes. Sole Mates is one of these firms (020 8524 2423) and Clarks also offers a service, so always enquire before you buy unnecessarily. It is sometimes possible to get special shoes on prescription which can be arranged by your specialist and the surgical appliances department.

If your child's feet are very slim as well as small, it may be worth trying sports trainers which are made in very small sizes (from a size 2); they also help support the ankles.

MEDICAL FEATURES REQUIRING TREATMENT

Hypoglycaemia

As previously described, low levels of sugar in the blood may be a problem over the first few months of life and in the most extreme cases may require tube feeding, particularly overnight. **Maintaining a constant calorie intake and avoiding prolonged periods without food, particularly during illnesses, is probably the most important thing to encourage.** In addition, if your child does have regular problems with hypoglycaemia, your doctor may recommend the use of a glucose gel which can be rubbed inside the mouth.

Emergency treatment for recurrent hypoglycaemia may be needed and you should get advice from your specialist. In addition, it is important that you have good access to your GP and, if necessary, **the nearest Accident & Emergency department** of a hospital (NB: this may not be at the nearest hospital and you should check this with your GP).

Surgery

Particular care should be taken if your child requires an operation as this may involve a prolonged period without food before, during and after the operation. Your doctor will advise as to what should be done and it may be that your child needs to have glucose given intravenously to cover the period of the operation.

In addition to the aspects mentioned above, if your child is to have an operation it is important that the anaesthetist is aware that your child may be physically smaller than their age would suggest and so will probably require less anaesthetic. Normally, the dose would be calculated on the basis of the child's weight, and this would be measured on the ward, but there is no harm in checking that this has been done.

Puberty

Puberty, and the associated physical changes, usually occurs at the appropriate age and progresses normally. In a few children it may start slightly early but still within the expected age range.

GROWTH AND GROWTH HORMONE INSUFFICIENCY

Much research indicates that between 10% and 30% of children with low birthweight or Russell Silver syndrome have abnormalities of growth hormone secretion and it is in these cases that the use of growth hormone (GH) has a stronger indication as a treatment. However, the majority of these children do not have an insufficiency of growth hormone. The use of growth hormone to increase the short term growth rate has been established, whereas evidence as to the outcome of final height is still awaited. However, recent trials have been promising and growth hormone treatment may help improve growth in some children with IUGR/Russell Silver syndrome.

The growth response to growth hormone treatment is variable. However, in those children who have benefited it appears that improving their growth rate creates a great boost to esteem and confidence and this can be for a number of reasons. Even if the eventual adult height is not significantly improved, allowing a child's height to be in keeping with their peer group, especially as they progress through school, may be beneficial. Also, if the child is receiving growth hormone there may be an increase in food intake. Although not necessarily putting on weight, there is sometimes an increase of muscle tone and parents report that their child appears more 'robust' and apparently seems more able to fight off infections.

SUMMARY

The term IUGR probably represents a spectrum of conditions (some of which are described as Russell Silver syndrome) resulting from abnormal foetal growth. As with any syndrome, not every child will have all the features described. The child who has IUGR, but has not experienced 'catch-up' growth during the first year of life, will remain small for their age and probably very thin. Their final height may be in the region of 157 cm (5' 2") for a boy and 144 cm (4' 9") for a girl.

There are many features of IUGR which may require medical help and support and it is important that you discuss any concerns that you have with your growth specialist who can then refer your child to another appropriate specialist if necessary. This may be especially relevant when considering leg asymmetry and the referral to an orthopaedic surgeon with experience in the procedure of limb lengthening (and not shortening).

There is no definitive way to help with achieving weight gain although it often causes much concern. These children are healthy and active, and confrontation over food

should be avoided if possible. There is no treatment at present that can be of certain long-term benefit for growth. The use of growth hormone appears to improve the rate of growth in the first five years of treatment, but longer-term benefit is as yet uncertain and clinical studies are still ongoing. The estimates of adult height after growth hormone treatment will not be known until these studies are complete.

If both parents are healthy and show no signs of the condition of Russell Silver syndrome, the risk of a further child being affected is very small (about 5%). However, genetic research is continuing to help try and understand more about the condition and the causes.

We hope that this booklet has helped you to understand more about your child's condition. While we have tried to cover the most important features of this syndrome, it must be stressed that very few children have more than a few of the characteristics which we have described.

QUESTIONS AND ANSWERS

Q1 Does the sweating seen in children with Russell Silver syndrome/IUGR improve as they get older?

A The most common characteristics of this syndrome are the poor appetite and the sweating. The sweating usually occurs at night and some parents have described picking up their child out of bed and both them and their bedclothes being soaking wet. This does appear to improve as they get older.

Q2 Does their appetite improve as they get older?

A Yes, but not completely. When children with Russell Silver syndrome enter puberty there is usually an improvement in their appetite but it still remains far from normal for someone of their age. These children usually remain thin.

Q3 Can the diagnosis of Russell Silver syndrome be confused with other conditions?

A Yes. As there are very few signs and symptoms which are completely specific to Russell Silver syndrome, it can be difficult to confirm the diagnosis. Unfortunately, at the moment, there is no blood test which confirms the diagnosis although this is being studied. So, the diagnosis needs to be made by an expert in this condition and this is usually an endocrinologist or a geneticist.

Q4 Is the body asymmetry more often right- or left-sided and does it alter with age?

A It is not known whether there is more right- or left-sided asymmetry. However, it can appear that the left side is more often smaller than the right. It is known that the difference remains unaltered both as the child gets older, and in adulthood.

Q5 Does growth hormone treatment improve final height in Russell Silver syndrome/IUGR?

A This question has not yet been answered by the research that is being done. Certainly, the dose that is used in children with growth hormone deficiency has very little effect in children with Russell Silver syndrome. It seems that a dose about twice as high as the normal 'replacement' dose is needed. In studies looking at these higher doses of growth hormone there has been a short-term improvement in growth rate but the results are confusing because there is no change in the predicted final height of the children being treated. Studies which go on for longer, that is until the children being treated reach their adult height, will be needed to see if the treatment helps them to be taller than they would have been without treatment.

Q6 Is growth hormone therapy helpful in treating the body asymmetry?

A No. The body grows entirely normally and both sides of the body respond equally and so the asymmetry remains.

Q7 Should children with Russell Silver syndrome/IUGR who also have growth hormone insufficiency be treated with growth hormone?

A Yes. This is a positive indication for growth hormone treatment. Also, growth hormone treatment tends to raise the level of sugar in the blood and so can be helpful in preventing hypoglycaemia, especially at night.

Q8 Is special care required in children with Russell Silver syndrome/IUGR if they are to have an operation and an anaesthetic?

A Yes. It is important that the parents explain to the doctors and nurses that a long time without food (sugar) is very dangerous in children with Russell Silver syndrome/IUGR. Great care should be taken if prolonged starvation, such as can be the case before an operation, is needed. These children are more likely to develop hypoglycaemia because they have very limited store of sugar in the liver. If there is any concern, a glucose infusion (a drip) should be set up before the operation and should be continued during the operation as well as during the recovery period. Regular checks should be made of the level of sugar in the blood.

Q9 Do children with Russell Silver syndrome/IUGR live to a normal old age?

A This is a difficult question. There is evidence that having a very low birth weight can lead to an individual having an increased risk of heart problems. However, this is based on people who were born over 40 years ago and so whether it is true for children born today will not be known for a further 40–50 years.

