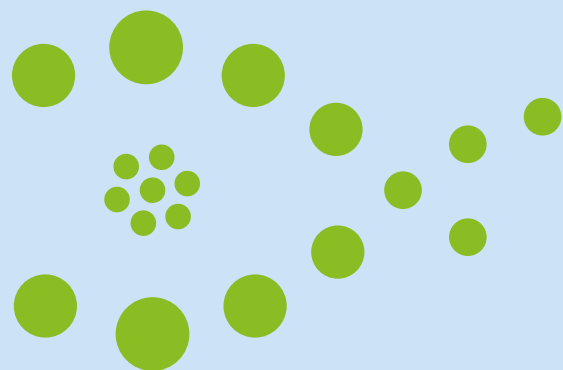


Booklet for Schools and Colleges

supporting children and young people
with Bardet-Biedl syndrome in a
learning environment





This booklet is based on the views of children and young people who have BBS and their parents and carers and has been written in collaboration with medical and education professionals, experienced and interested in BBS. Our aim is to provide a greater awareness and understanding of BBS for those working with children and young people who have the syndrome, to enable them to more effectively support their students to reach their full potential.

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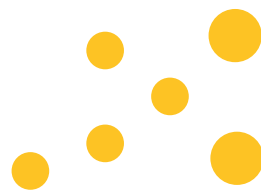
An Introduction to Bardet-Biedl Syndrome (BBS)

Bardet-Biedl Syndrome (BBS) is a rare genetic disorder. Features of the syndrome include:

- Rod-cone dystrophy, a progressive eye disorder that leads to loss of vision, characterised by tunnel vision and night blindness
- Obesity
- Renal abnormalities
- Developmental delay
- Emotional immaturity
- Speech and language difficulties
- Extra fingers and/or toes
- Learning difficulties

Not all the features listed are always present in those diagnosed with BBS and each one can vary in severity and appearance, so it is important that all sections of this booklet are considered to fully understand BBS and the implications for children and young people living with the syndrome.

Key Action: Student Passport



At the back of this booklet is a section for the child/young person to complete describing how BBS affects them. This should form the basis of a Student Passport, a concise but comprehensive document, readily available to all staff working with the student to support learning, teaching and access to the curriculum and learning environment. By focusing on specific provisions and considerations that need to be made, the Student Passport gives the student confidence that people are aware of their needs and appropriate strategies are being shared.

A Student Passport template can be downloaded from the BBS UK website: www.bbsuk.org.uk under 'education'.

We asked students, 'what do you think your teachers need to know about BBS?'

To understand my type of vision problems, to understand this can affect my emotions, to understand my low pain threshold.

They need to know all about the syndrome and need to know how to keep me safe.

They need to actually know what it is, none of them understand how it affects me.

Rod-Cone Dystrophy

One of the main features of BBS is rod-cone dystrophy. It usually begins as night blindness during primary school years and is a progressive degeneration of the retina that leads to a restriction of the visual field and, in most cases, a registration of severe sight impairment, usually during secondary school years. Visual impairment may not be detected during the early school years as fragmented vision can be masked by the young person who is unaware that there is a problem and/or where there are speech and language difficulties. A sensory team or qualified teacher of vision impairment (QTVI) should be involved from an early stage.

The student's vision may deteriorate throughout the school years and fluctuate throughout the school day depending on light and energy levels. An individual with a static visual impairment will learn their limitations and develop adaptive coping strategies, however when the visual impairment is progressive and fluctuating, these strategies will be less effective and protective. A student with rod-cone dystrophy will experience vision fatigue and staff members should be sensitive to this. A Student Passport would be useful to highlight risk and ensure safety within the environment.



The following pages in this section provide useful strategies for anyone working with someone with a visual impairment. Thank you to Stockport's Sensory Support Service for this advice.

Tips on vision friendly environments

- Keep the layout of the classroom the same, keep major changes to a minimum.
- Remove obstacles and furniture that may cause an obstruction and/or restrict available space.
- Floors should be kept clutter free, chairs should be tucked in.
- A visually impaired student's cloakroom peg will be easier to find if located on the beginning or end of a rail, rather than in the middle.
- Keep lighting bright, low lighting will make it more difficult for those with BBS to use residual sight.
- The light source from the windows should come from behind, or to the side and glare can be avoided by adjusting seating whenever necessary. Being able to control light is important e.g. blinds may need to be used on particularly bright and sunny days to reduce glare.
- To present displays, use clear print, in a large accessible and simple font e.g. Arial or Comic Sans and keep layout simple and logical to follow.
- Present displays on matt, well contrasted backgrounds, at a height from which children can easily read, avoiding shiny surfaces.
- Use matt lamination (not shiny) to protect displays, flash cards and activities.
- Provide additional space at the child's desk for equipment and large print documents and additional storage space for folders and equipment in the classroom if needed.

Not always having my resources for the lesson prepared beforehand means I can't always do as much as everyone else.

Student

Tips on vision friendly teaching approaches

- Stand away from the window when teaching or speaking to the children, to prevent creating a silhouette, particularly on a bright day.
- Position the desks and chairs so that where possible, there are opportunities for children to be positioned facing and near to the front.
- Verbalise what is being written on the board. Point to the text on the board as it is spoken about.
- Print in large clear writing, using a black pen on the board. Keep layouts simple and logical to follow.
- Clean class and individual whiteboards regularly to aid contrast and clarity.



Sometimes I can't see stuff, so I can struggle in the lesson. When they turn the lights off to use the board it's frustrating as I can't then see because of my [poor] night vision.

Student

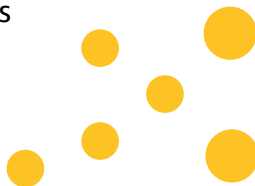
- When using the smart board, make adaptations where possible to optimise access, e.g. text in black, large print size, simple and logical layout.
- Adjusting to different light levels takes longer in those with rod-cone dystrophy and staff should try to avoid repeatedly switching the classroom lights on and off.
- Recognition of facial expressions from the teacher may be poor. Speak to all children by name when they are taking part in class discussions, or when they are answering questions.

Printed materials

- Table top copies of board work should be routinely provided at the start of all lessons for easier access.
- Keep the layout on worksheets simple, with good contrast against a plain background avoiding visual clutter, overlaid text and small detail.
- Use A4 size paper to present worksheets.
- Use a simple font (Arial or Comic Sans) no smaller than size 14 font as standard. Those with a restricted field of vision will not necessarily benefit from enlargements.
- Ensure diagrams and pictures are at an accessible size and not too small to distinguish detail. They should be presented with clear black outlines and carry clear labelling.
- Avoid text over the top of a picture.
- Avoid situations where worksheets/books need to be shared.

Technology

- Learning to touch-type should be commenced as soon as possible.
- The introduction of braille may be appropriate during late primary/secondary school years.
- An assessment by a qualified teacher of vision impairment (QTVI) may identify additional useful equipment and strategies.
- For older students, access to a laptop/tablet can be beneficial allowing the young person to enlarge documents and adjust contrast independently as needed. Lesson resources should be pre-loaded or emailed in advance to the student.



Tried and Tested

"Braille is being introduced and taught at the student's own pace to develop literacy and numeracy skills. We hope this will lessen the inevitable emotional impact of future sight loss."

"We use enlarged font text and an enlarged computer keyboard to support our student's learning, but also use a portable magnifier to enable them to read smaller text and examine objects in more detail."



Useful Website:

www.rnib.org.uk

Moving Around

- Lighting should be kept bright, constant, and without shadow in all areas of the school, including corridors, cloakroom areas and toilets.
- Steps around the school and changes in surface levels should be edged with yellow paint or striped tape and have handrails fitted where appropriate.
- Corridors should be clear and not have items stored alongside walls, such as chairs, propped up display boards and so on.
- Staff should be aware that after leaving a bright playground, the eyes of a child with Rod-Cone Dystrophy will take extra time to adjust to dimmer lighting indoors.
- Tinted lenses are sometimes worn, and care should be taken in dim lighting that safety is not endangered.
- Supervision should always be provided for young visually impaired students in new or poorly lit environments and on school trips.
- Some children and young people would benefit from mobility training at an early age.

Tried and Tested

"We are fortunate to be visited regularly by an effective VI team who offer support to both the staff and the young visually impaired student."

Useful Website:

www.guidedogs.org.uk/services/children-and-young-peoples-services

Social Interaction

Many children and young people who have BBS struggle to make and maintain friendships. Social interaction opportunities are vitally important from a young age to enable young students who have BBS to develop their social skills, ahead of any potential visual deterioration. Proactive strategies and support, including attendance at after school clubs, should be encouraged and included in the child's individual education plan (IEP) or education, health and care (EHC) plan. Parents may appreciate support with identifying friendships to develop outside of school.

Children and young people with BBS may find interpreting non-verbal communication difficult and may respond in a way that appears inappropriate. This can lead to confrontation and conflict, which can be confusing to the young person who is unaware of the part they played in the conflict. Support for the whole peer group, perhaps within the PSHE aspect of the curriculum, would be beneficial.

The young person may struggle to find their friends in a busy playground and strategies should be put in place to make this easier. Consideration should be given to providing sighted guide training to the friends of the visually impaired young person, so they can assist where needed enabling greater independence.

Confidence building exercises benefit everyone and subjects such as drama or dance can help with social interaction and integration.



The school playground can be a confusing place to play. So many children all dressed the same and moving around so quickly can cause many obstacles for a child with a visual impairment. Just finding their friends can become a frustrating task.

Parent



My friends have had mobility training which has helped if I need support from them. This helps me to have some independence without adults.

Student

Sports

It is important that all efforts are made to include the student in the activity. Support may be needed for safe inclusion in many aspects of PE and the understanding of fellow pupils will be vital to encourage peer support. Gym sessions and swimming are all accessible and some sports are more easily adapted to suit the needs of a visually impaired person than others.

Difficulties often arise during PE and ball games as the young person's vision deteriorates. A restricted field of vision makes it difficult to keep track of a rapidly moving object whether that be other people, balls or equipment.

Specialist equipment can provide easier access, e.g. yellow balls or balls with sound for football, netball, cricket or hockey.





I'm really into sports and when I found out I had BBS, I didn't think playing sports would be possible. However, a whole new world opened up for me after I went along to a vision impaired cricket event. I won the best new player award at my first match.

Former student

Children and young people who have BBS often struggle to maintain a healthy weight which increases the importance of physical exercise, although this can be challenging and hold the young person back. Orthopaedic abnormalities and poor balance and co-ordination can also feature in BBS and many have a wide gait, which can significantly impact on the young person's sporting ability.

Young people who have BBS can also struggle with fatigue and may take longer to recover from a sporting activity than their peers.



Useful Websites:

www.britishblindsport.org.uk

www.metroblindsport.org

www.rnib.org.uk

Learning Difficulties and Developmental Delay



There is a great variation in learning difficulties among the BBS population, with some individuals seemingly unaffected by learning/behavioural difficulties whilst others can have global developmental delay and/or are diagnosed on the autistic spectrum. Studies conclude that a significant number of individuals with BBS experience significant learning difficulties, but only a minority show a severe impairment in IQ tests.

Developmental delay is common in children who have BBS, including speech and language development, general motor skills and, to a lesser extent, fine motor skills. Some children and young people may take longer to respond to a question, however once decoded, the question is mainly answered in a reasoned and intelligent way. The 'delayed response' phenomenon is widely reported by parents and teachers of children who have BBS.

Some common difficulties for children with BBS are:

- Emotional immaturity
- Poor reasoning
- Poor communication
- Psychological inflexibility
- Obsessional thinking
- Rigid routines
- Challenging behaviour
- An inability to form friendships

An EHC plan may be beneficial to ensure that a child/young person is fully supported throughout their education and an assessment should be undertaken where appropriate. It is important to get any associated learning difficulties such as autism diagnosed, as a diagnosis could then lead to help from other areas. Depending on the specific needs of the child/young person, an EHC plan will contain recommendations from appropriate professionals such as:

- Educational psychologist
- Speech and Language Therapist
- Sensory Support
- Learning Support
- Behaviour Support
- Physiotherapist
- Autistic Spectrum Disorder/Condition Team
- Medical personnel
- Occupational Therapist (can advise on helpful specialist equipment such as a sloping board, grips for pens, scissor types etc. to help develop fine and gross motor skills)

Tried and Tested

"We find that what really works is clear and concise communication at an appropriate word level, with plenty of time allowed for the student to process what has been said or asked of them."

"The student I support responds well to clear boundaries and expectations. Each morning I give a brief overview of the day, so they are aware of any likely changes, which they may find difficult."

Useful Resources:

The Motor Skills United Occupational Therapy Programme is a useful resource that can be implemented in schools by teaching assistants; it can be purchased from www.specialdirect.com

Emotional Difficulties in BBS

Between the ages of 7 to 12, there is typically an increasing awareness of the condition and a feeling of being different starts to emerge, particularly in a school setting. At this sensitive age, young people may have difficulty with friendships, obesity and bullying, resulting in low self-esteem.



For some children, much of this improves as they mature; however, emotional immaturity, poor reasoning, inflexibility and obsessional thinking can carry on into adulthood. Depression, anxiety, panic attacks, anger and poor emotional control commonly affect young people with BBS. It is unclear whether this is a part of the syndrome or an indirect result of it. Most likely, it is probably a combination of both.

Many parents report that their children strongly dislike loud noise and disruptive classrooms and can quickly become distressed and tearful. Sudden noisy situations such as a fire drill can be very upsetting and uncomfortable, and the child/young person should be forewarned and supported if needed.

Someone with BBS is likely to take everything that is said literally and to heart, which can lead to worry and stress. Words should be chosen carefully, and exaggeration avoided. The behaviour of other class members may sometimes warrant a stern and direct approach but facial recognition difficulties and visual impairment in general can mean that a child with BBS may not realise that the teacher's disapproval is not directed at them, so attention should be given to the impact this can have.

Changes of routine and environment can cause distress and anxiety and so it is essential to take the time to discuss what is going to happen and familiarise the young person with the changes e.g. visitors to the classroom, school trips and school events.

Making the transition from one class/year group to another can be stressful, but this can be eased by working with the child beforehand. Being made aware of new routines and a new environment will help to increase confidence, e.g. practising different routes to places around the school, familiarisation of the layout of a new classroom and identifying the location of where to store things such as coat and bag. Be mindful of little things that could make a difference such as having a coat peg at the end of the row rather than in the middle. The child should be encouraged to discuss the things they may be worried about so that they aren't left worrying (possibly over the school holidays) about how they are going to cope with their new challenges.



Routines are a part of school life but when change is involved – for example, new classrooms, new teachers, and new cloakroom areas - where possible, these changes should be frequently discussed in advance, so as not to cause too much worry and disruption. More time should be taken to introduce and familiarise children with these changes.

Parent

Useful Websites:

www.autism.org.uk

www.autismeducationtrust.org.uk

Challenging Behaviour

Build up a useful bank of strategies:

- Avoid confrontation. Discuss and deal with any issues only when the child/young person is calm.
- Find appropriate ways to distract the child to get the situation under control.
- Try and counteract any disappointments with something positive.
- Give warnings about changes in routine or events that are going to happen.
- Give the child a finishing time to work towards so they have some control of how to pace/organise themselves. Maybe make use of an egg timer to give some guidance as to how long they need to spend on a task or to indicate how long before the activity needs to come to an end.
- Have a visual reward chart with stickers and give the child opportunities to make up for disappointing behaviour.
- Create opportunities for the child to have some time out of the classroom if the environment becomes too stressful in some way e.g. increased noise level.
- Have a visual timetable available.
- Use a 'first and then'/'now and next' board.
- Make use of social stories.



Tried and Tested

"We find a visual timetable works well. We offer a small list of choices so that the young person feels in control and can build independence and confidence. We offer differentiated work where possible to enable the young person to feel able to achieve what is expected of them."

Useful Resources:

Circle of Friends can be a valuable strategy to reduce the social isolation of a young person and promote inclusion in a school environment. For more information go to www.inclusive-solutions.com

Communication in print allows everyone fair and equal access to information regardless of their level of language understanding and can be used to produce social stories for younger children and/or children/young people with autistic spectrum disorder. For more information go to www.widgit.com.





Speech and Language

Many children with BBS are affected by speech and language difficulties, which may range from mild to severe and can create a barrier between a child and the teacher/peer group.

Below are some examples of the types of difficulties that can occur:

- Expressive language impairment can include difficulties with putting sentences together, describing a sequence of events, telling a story and slow acquisition of new words.
- Difficulties with receptive language can affect understanding words and sentences. It can also affect following stories and more abstract language within the classroom and at home.
- Impaired social communication skills; some children may have a diagnosis of autism.
- Speech and sound development can be delayed.

A referral to a speech and language therapist should be made at the first signs of any speech and language difficulties so that appropriate strategies can be provided and put in place. This provision may be included in an EHC plan or IEP.

The use of Makaton can be a helpful tool for those struggling to communicate. All the class can be involved in learning the basics of communicating in this way.

Useful Resources:

ELKLAN interventions have been used successfully by teachers of BBS students; further information and training can be obtained at www.elklan.co.uk

Useful Websites:

www.talkingpoint.org.uk
www.ican.org.uk

Obesity

Obesity commonly affects those with BBS, usually commencing in childhood and increasing in severity with age. The cause of the obesity associated with BBS is unknown but is the subject of current research. There is no single treatment approach, however those with BBS can successfully lose weight and a healthy well balanced diet and active lifestyle approach is advocated.

Obesity in BBS exposes children and young people to bullying and it is vitally important that the teaching staff and support team understand that being overweight is not always because of over-eating and that they communicate this to the other members of the class and school, integrating it within their anti-bullying programme.

Many schools have signed up to a Healthy Schools Programme and supporting a BBS student's dietary needs will be considerably easier within this framework. Additional support may be needed but this should be as unobtrusive and sensitive as possible. If there are plans to offer unhealthy or additional foods during a lesson or school day, this should be discussed with the parents of younger children or the young person and their parents and a healthy alternative arranged if required.



Additional Features



Polydactyly (extra fingers and or toes)

Brachydactyly (shortness of fingers or toes)

Many babies born with BBS have extra fingers and/or toes. Although these will usually have been removed by school age, the removal of the extra digit often interferes with the structure of the hand or foot and can impair balance and dexterity. Shorter fingers can make everyday tasks such as the tying of shoe laces difficult and may pose problems with activities requiring fine dexterity and good fine motor skills. The ability to use keyboards or braille may be affected.

Neurological and Orthopaedic Abnormalities

Impaired co-ordination, balance and speech affects a significant number of individuals and low muscle tone is common amongst those with BBS of all ages. Many are also affected by hypertonia, a condition marked by an abnormal increase in muscle tension and a reduced ability of a muscle to stretch, which can cause significant discomfort.

Some individuals may also be affected by curvature of the spine, knock knee, bow leg and flat foot. Painful legs and feet can occur and orthotic intervention in the form of supportive footwear/ insoles is often prescribed to protect/support the foot and ease discomfort. These should be worn during PE to protect feet against further damage. Physiotherapy can also be beneficial and may be integrated into a PE programme and included in an EHC plan.

Support with adjustment of clothing and shoes may be needed before and after PE. Strategies for improved independence may include alternative uniform options, i.e. swapping a formal shirt for a polo shirt.

Kidney Abnormalities

Although Bardet-Biedl Syndrome can affect the kidneys in a number of ways, significant kidney problems and kidney failure are seen in only a small number of those with BBS. Several young people with BBS have experienced renal failure and undergone successful kidney transplants at primary and secondary school age. Children and young people who have chronic kidney disease will tire easily and may need to follow a special diet. A more common problem with the kidneys in BBS is a reduced capacity to concentrate the urine, which can mean increased urine output and sometimes excessive thirst. Children should have access to water at their desk and a relaxed attitude should be adopted by all staff regarding toilet access.

Hearing

Conductive hearing loss affects many children with BBS. It is almost always caused by glue ear which is common to many children, so most teachers will be familiar with how it can adversely affect the learning process. A small proportion of children also have sensorineural deafness.

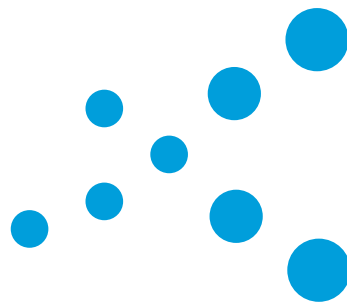
Useful Websites:

Sense is a national charity that supports people who are deafblind, have sensory impairments or complex needs, to enjoy more independent lives: www.sense.org.uk

RADAR Keys enable access to disabled toilets across the UK; to purchase a key go to: www.disabilityrightsuk.org/shop/official-and-only-genuine-radar-key

If standard disabled toilets are not accessible or sufficient for an individual's needs, Changing Places have more space and equipment, including a height adjustable changing bench and a hoist. To find a Changing Places toilet, go to: www.changingplaces.uktoiletmap.org

Puberty and Development



Urological and reproductive systems may be affected in both boys and girls who have BBS and puberty can be delayed for both. Privacy should be made available in the form of access to a separate/disabled toilet where appropriate.

Both boys and girls would benefit from sensitive counselling during puberty. This can be a difficult time when a young person's self-esteem may be low, and they may need help in coming to terms with all aspects of the syndrome. Self-image is particularly important because it reflects not only self-assessment of successes and failures, but also the attitudes that other people have shown them.





Further Education and Employment

Many young people with BBS have gone through or are currently in further education, with many studying successfully at college and university level. Some have enjoyed residential placements at specialist colleges, and others have gained employment or started their own businesses. It is crucial that young people with BBS are given every opportunity and the right support to achieve the future they are entitled to and are capable of achieving.

Supported Internships

A supported internship is one type of study programme specifically aimed at young people aged 16 to 24 who have a Statement of Special Educational Needs, a Learning Difficulty Assessment, or an EHC plan, who want to move into employment and need extra support to do so. Supported internships are structured study programmes based primarily with an employer. They are intended to enable young people with learning difficulties and/or disabilities to achieve sustainable, paid employment by equipping them with the skills they need for work through learning in the workplace. Internships normally last for a year and include unpaid work placements of at least six months. A useful fact sheet about Supported Internships can be found on the Preparing For Adulthood website: www.preparingforadulthood.org.uk.

Specialist Schools and Colleges

There are many specialist schools and colleges spread around the UK offering day and residential placements for students to follow academic, vocational and independent living skills courses. Those attended by students with BBS include:

- Royal National College for the Blind, Hereford
- New College Worcester
- Queen Alexander College, Birmingham
- WESC Foundation, Specialist Centre for Visual Impairment, Exeter
- Henshaws College, Harrogate

For further information and to explore other options, go to www.natspec.org.uk. Natspec is the membership association for organisations which offer specialist further education and training for students with learning difficulties and/or disabilities. The website enables users to search for specialist provision by location and expertise.



When I was 16 I went to the West of England College in Exeter. At the college I learnt how to be independent, doing such things as cooking, cleaning and shopping. I also completed courses in further education, including an NVQ in administration. I also did work experience placements. I love living independently and being able to see my friends and go where I want to. I feel that I have achieved a lot so far in my life, and I look forward to enjoying life to the full.

Former student



I have met so many children, young people and adults who have BBS at conference and clinics over the years and their bravery, courage and determination in the face of disability and illness is inspiring. I have also seen so much achievement within our group, whatever their passion; we have successes in business, sport, in the workplace, music, art, as well as academic and there is a great deal that can be achieved.

Clinics Support Worker



All About Me

Name

How BBS affects me...

How BBS affects me at school...

I find it difficult to...

It would help me if you could...

I will help myself by...

Additional Comments...

All About Me



Bardet-Biedl Syndrome UK

This booklet was produced by Bardet-Biedl Syndrome UK (BBS UK), the only UK charity providing support and information services to those diagnosed with BBS, their families and carers. Further information can be found on the BBS UK website or via the contact points below. BBS UK would like to extend their most heartfelt thanks to Genetic Disorders UK for funding this booklet via the Jeans for Genes Grant Programme. Our grateful thanks also go to Gareth D Morewood (www.gdmorewood.com), Director of Curriculum Support at a High School in Stockport for allowing BBS UK to adapt his original Student Passport.

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