TEACHER NOTES

A. Causes and effects of congenital deafblindness

Learning outcome 1: Understand the range, causes and implications of congenital deafblindness.

Many and varied causes; often no causes identified at all (resultant problems for parents, etc).
NB – important to understand the difference between *genetic* and *congenital.*

- Genetic causes.
- Problems relating to pregnancy/birth.
- Conditions with gradual onset.
- Causes with similar effects.

Genetic causes of deafblindness

Long list of conditions, many very minor.

Significant conditions:

- **Charge syndrome**
  Charge was previously an acronym where each initial stood for one condition associated with the syndrome but this is no longer the diagnostic criteria. People with Charge Syndrome usually have:

  – Coloboma (an incomplete eye in retina or iris)
  – unusual ears – including mild-profound deafness
  – heart problems, balance problems (organs of balance damaged or not existing)
  – problems with mouth and nose – cleft palate/incomplete tubing in mouth/nose for breathing and swallowing, limited or no sense of smell/taste
  – learning disabilities
  – short stature, genital underdevelopment (most obvious in boys).

- **Goldenhar syndrome**

  Malformations or abnormalities of facial structures, ears and eyes, often associated with brain damage, cortical visual impairment, and deafness. In addition, abnormalities often involve the skeletal, cardiac, central nervous and renal systems. Hearing loss varies from near normal to severe; vision defect, including diplopia of various degrees. Moderate learning disabilities may occur in about 10% of cases.

- **Moebius syndrome**

  Problems with facial muscles, hearing, visual and speech impairment.

- **Refsum's syndrome**

  Problems initially of unsteadiness and/or failing vision, leading to night blindness, progressive (nerve) deafness, loss of sense of smell, unsteady gait, etc.

Deafblindness caused by problems in pregnancy/birth

Many possible problems, including:

- **Congenital rubella syndrome**

  Although rubella is not particularly serious for children or adults, or for the pregnant woman herself, it can seriously damage a baby's organs in the early stages of pregnancy. Some of the common effects include impairments to the:

  – ears: a child may have hearing loss in one or both ears due to damage to the inner ear, which links the ears to the brain
  – eyes: babies may be born with cataracts (cloudiness to the lens) in one or both eyes. Others may have rarer visual conditions or find that their sight gets worse as they get older
  – heart: rubella can affect the heart in many ways. Children may have heart problems from birth and require hospital treatment
  – brain: rubella can also affect a child's brain and nervous system. Difficulties can vary from mild to severe.

- **Prematurity**

  Premature infants may have incomplete ears or damage caused by strong drugs against infection, or damage to eyes (retinopathy of prematurity) because of oxygen delivered in

incubator. Retinopathy of prematurity can lead to no vision, or blotchy vision (with areas of retina which don't work properly).

- **Birth asphyxia**
  Generalised brain damage (including severe/profound learning disability), motor (movement) problems, slow mental development, seizures, poor hearing or deafness, poor vision.

Deafblindness caused by deteriorating conditions

Hearing and/or vision may be within or almost within typical limits at birth but then deteriorate. Some conditions (e.g. metabolic diseases) often have a poor outcome; deterioration of vision and hearing, and difficulties thus caused include general severe deterioration. Other conditions have deterioration only of vision/hearing.

- Metabolic diseases, e.g. **Hurler's syndrome**
  Poor outcome for many – vision deteriorates around age five, usually to blindness, hearing begins to deteriorate at same age but may continue useful for longer, gradual deterioration in cognitive function and early death.

- **Alstrom's syndrome**
  Early visual impairment often severe, hearing gradually deteriorates in teenage years, other problems may occur such as diabetes.

- **Usher syndrome**
  There are three main types of Usher:

  – Type1: born deaf, serious visual deterioration due to Retinitis Pigmentosa (RP) around adolescence, balance problems, maybe as many as 1 in 20 profoundly deaf children.
  – Type 2: moderate severe deafness from birth and deteriorating vision (RP) around 20 (or before).
  – Type 3: vision (RP) and hearing deteriorate simultaneously (or very mild early hearing impairment) often in middle age.

  Retinitis Pigmentosa: name for a group of hereditary diseases of the retina. The retina is the light sensitive tissue at the back of the eye where the first stages of `seeing' take place. With Retinitis Pigmentosa the retina slowly degenerates and loses its ability to transmit pictures to the brain. The early symptoms of Retinitis Pigmentosa include difficulty seeing in

the dark, often coupled with problems in adapting to bright light and changing light conditions. Loss of visual fields which may give a patchy effect to side vision is also common.

Tunnel vision: in one type of retinal degeneration the cells at the edge of the retina gradually stop sending information about changes in light levels and the shape of objects on the edge of someone's vision. This loss of visual field, or `tunnel vision' means that someone cannot see objects unless they are directly in front of them. This may mean for example, that they can still read the bus timetable, but will need help to find the bus station.

Loss of central vision: another type of retinal degeneration causes the cone cells in the retina to be affected first so that the individual can no longer see the details of the object in front of him and will find it increasingly difficult to read print.

Obvious emotional and adjustment difficulties of Retinitis Pigmentosa.

Conditions which cause similar effects but which may not be existing at birth

- Early meningitis.
- Non-accidental injury – which can damage brain/visual/hearing mechanisms and lead to similar effects.

Range of deafblindness/dual sensory impairment

Most congenitally deafblind people have residual vision and/or hearing – 4% of deafblind children may have minimal or no vision/hearing.

The existence of some vision and/or hearing, which may be distorted, from birth, does not mean that the person is not deafblind – they have poor access to information, difficulty in typical communication, and with mobility. Vision and hearing usually work together, or one can compensate and give access where the other is limited, in dual sensory impairment this often cannot happen.

Visual impairment is part of a continuum, with some (very few) being completely blind. The ability to see only light can help a person move around. Others may have vision for some tasks (e.g. looking at things close to them, or reading) but not others (cannot move around as field of

vision is limited). Some will manage well where an environment is adapted, e.g. kitchen with dark surfaces and light utensils, but not where they are not).

Combination of vision and hearing impairment by itself can lead to a **delay** in:

- learning
- communication
- mobility and motor skills
- social and independence skills
- difficulty in behaviour.

Some individuals may have additional **disabilities** in the following areas:

- brain damage
- language difficulty
- cerebral palsy
- behaviour difficulties.

Some causes of congenital deafblindness affect other systems:

- Some congenitally deafblind people will have medical problems, e.g. heart (e.g. Charge, Rubella) or feeding, breathing (prematurity).

These additional difficulties can cause further problems with access to the environment and learning, for example:

- Mobility problems (e.g. cerebral palsy or motor dysfunction) which can make exploration in space, or with hands difficult.
- Balance issues (e.g. Charge syndrome) which can make walking slower to develop.
- Slow concept development, generalised learning disability making it harder to connect perception.
- Lack of access to communication can cause frustration and emotional difficulties (particularly in the case where deterioration of senses occurs).

Congenital dual sensory impairment affects the whole life experience. High expectations are needed to ensure potential is achieved; because of a range of difficulties, development may be delayed, but this does not mean it's impossible.

Dual sensory impairment may cause delays in developing communication skills, learning concepts, social skills, and problems in managing behaviour, but this may not be due to additional disabilities. Learning disability or motor disability in particular should not be assumed.

Congenitally deafblind people don't suddenly become like adults with acquired deafblindness when they reach adulthood – their whole life experience is different and levels of achievement may be different.

B. Provision and roles

Learning outcome 2: Know the roles of people who can help promote and develop communication with congenitally deafblind people

- Specialist teacher
  A qualified teacher of children with deafblindness (sometimes called MSI – multi-sensory impairment – in education) should be working with a dual sensory impaired child in a class teacher or advisory capacity.

  The teacher should be able to advise on assessment of communication levels, individual plans for developing communication, and alternative and augmentative communication (signs, symbols, cues, etc).

  They should be able to advise on environment, liaison with home/family etc and next steps.

- Intervenor
  An intervenor is a person who works one to one with a deafblind person. An intervenor is not simply an interpreter, they enable the deafblind person to have access to things beyond that person's ability.

  They may arrange transport and accompany the person on trips (usually in adult work).

  In schools, they are more likely to support delivery with tactile versions of lessons, appropriate communication, and alternative approaches.

  The intervenor does with, not for. The intervenor will promote independence and increased access. An intervenor should provide consistency in understanding early communication and providing correct cues. They may be key to developing a secure relationship for a child, enabling them to make sense of communication.

The intervenor may also be working with the family and in the community to facilitate access.

- Speech and language therapist
  A speech and language therapist may help with providing access to alternative and augmentative communication and assessment. Provides activities for access to communication and intensive interaction.

- Teacher of the deaf/visually impaired may also be involved with deafblind children.

C. The experience of congenitally deafblind people

Learning outcome 3: Understand the factors influencing the experience of congenitally deafblind people and their involvement in society.

Congenitally deafblind people have different life experiences from those who acquire deafblindness.

Life experience depends on factors such as:

- amount of vision and hearing
- stability in home background
- appropriate education as dual sensory impaired person

(and, where vision and hearing loss are progressive)

- emotional issues (grief over deteriorating senses).

Expectations and achievements

Expectations – people with congenital deafblindness may not necessarily have an additional learning disability, but low expectations can cause low achievement.

Achievements (e.g. for people with Charge syndrome) can be measured in terms of:

- going to university
- getting and enjoying a job
- getting married
- living with other people.

For others, achievement may be in terms of:

- amount of independent living
- having friends and a social life
- having interests and hobbies
- finding some means of being understood
- finding some activities which are enjoyed.

Communication needs and preferences

- It is important to remember that some deafblind people may prefer to live more private lives (just as those who are not dual sensory impaired).
- To enable communication, there must be something worth communicating,and or talking about, at whatever level, e.g. a trip out (at a more sophisticated level) or a great experience in the sensory room (fun with lights).
- Dual sensory impairment can mean that there is not enough to talk about or do. Experiences may have to be specially adapted so that they can be close enough to be understood/shared. They need to be distinct (what's the difference between shopping, and going to a museum, for a person with only immediate communication in context).
- Security is especially important for many deafblind people, for whom life can be random and disordered. This is accomplished by structure and routine, enabling deafblind people to build up relationships. However, life can be boring for everyone, so the need for novelty is also there – new experiences, different things.

The environment

- Environment should be adapted to ensure that a dual sensory impaired person is able to participate as fully as possible.

- Other people need to be willing to:

  – share the communication mode
  – give time to wait for response
  – listen to what is being said
  – understand.

- Congenitally deafblind people's communication is disrupted from birth, (from eye contact and response to sounds) – for some this may be very severe. This may mean they need a huge amount of help to build relationships at any level:
  - relationships can mean friendships with chat
  - relationships can mean the ability to share a laugh together, during a massage or when something is spilt.

- Lack of communication can lead to emotional and behavioural problems, and difficulties in managing behaviour, e.g. deteriorating vision, particularly if there isn't the language to express, understand or discuss it, can lead to behavioural problems.

- However, the structured, adapted environment should always link with the actual needs of the person:
  - provision of carefully tactually marked door-handles, with emphasis on always feeling them, would be of little value to a person who doesn't have formal communication, uses a wheelchair and doesn't understand much about buildings because of their level of concept development.
  - Little point in having symbols for 'Friday' and 'Monday' if the person has little understanding of today and tomorrow.

D. Independence and community

Learning outcome 4: Understand how to develop opportunities for maximising independence and communication for the congenitally deafblind person

- A congenitally deafblind person is likely to need support in communicating with the wider public (hence the importance of transparency – labelling symbols, etc).
- Asking for assistance is an important skill to be acquired: by simple tap and request – with symbol or help card, etc.
- More complex forms can be used, even if not understood, through the use of cards with descriptions e.g. 'Please can you help me get on the bus' or 'I want a sliced brown loaf please'.
- Where a symbolic level has been reached, independence can be fostered by use of lists and schedules – for getting ready for school/centre/work or for making a cup of tea, using photographs, symbols, or tactile symbols or printed words.

- For those who have not reached this level, the use of routines, and placement of objects (right to left for example, or in basket to out of basket) can help to give information and allow maximum independence.

Daily living tasks

- Adaptations for people with visual impairment often rely on sound – talking scales, talking microwave, etc.
- Safety issues. Some simple gadgets are available in the general market, e.g. safety grater and onion fork, kettle pourer, microwave with dial. Other specialist items are available.
- Important to encourage indications of preference for certain foods, on basis of taste/texture
- Careful choosing of clothes for tactile qualities.
- Mobility/health and safety: ability to travel about, use of cane or other mobility devices for independence, safe use of electrical appliances, etc.

Community activities

- Involvement with other people in community activities may be limited by other people's ability to accept and communicate.

- Suitability of activities for those with dual sensory impairment.

- Understand that there is almost nothing that can't be done, but some areas with more immediate appeal include:

  – Physical activity – swimming, tandem riding, walking, gym, etc. Ball games (likely to be more limited). Importance of exercise for those with limited mobility due to visual difficulties, as they can't move around without help.
  – Music – especially live, but also recorded – sound light environments/sensory rooms/multi-sensory environments.
  – Caring for animals.
  – Shopping (not being afraid to choose a bubble bath by taking the lids off and smelling them!).
  – Cooking, eating and eating out.
  – Parties and social gatherings (some may like a busy, noisy environment sometimes).
  – Religious activities.

- Dancing – disco, ballroom, etc.
- Trips out.

These are especially important to the deafblind person who may not otherwise have much to communicate about, and need the opportunity to have exciting experiences.

- Hobbies to pursue independently may be more limited and may need to be carefully arranged beforehand. Nothing is impossible, but most depend on vision/hearing which is available. Examples which need little adaptation:

  - Knitting.
  - Collections.
  - Cooking.
  - Computer programmes.
  - Listening to music.

E. Learning and communication needs of congenitally deafblind people

Learning Outcome 5: Understand the learning and communication needs of congenitally deafblind people.

NB It is useful for learners to understand the historical progression of support for congenitally deafblind people

- First class specifically for deafblind children was held in 1944.
- Early classes taught behaviourally, with language taught as stimulus/response.
- Greater understanding of communication developed with use of signing, but some people, especially those with rubella, seemed unable to acquire it (possibly because of motor dyspraxia).
- Understanding is growing: those who are not using formal language are seen as potential communicators.
- Recognition of presymbolic communication, leading to understanding of some people for the first time.
- Greater understanding of the need for intersubjectivity – communication is about sharing, and the need to develop turn taking, sharing activities. Communication as connection.
- In 1988 DfES recognised deafblindness as educationally significant. Now specialist profession of teachers of deafblind children.

- Deafblind children taught in a variety of places, communication strengths sometimes depend on where they are taught.
- Some specialist classes and units taught by specialist teachers of deafblind children.
- Some deafblind children work with intervenors in special or mainstream schools.
- Many deafblind children are in schools for children with severe learning difficulties (but their difficulties are not always recognised).
- Some deafblind children have help (intervention/interpretation) in schools for the deaf/visually impaired. This may affect their communication style.
- Adults may be in specialist provision (often Sense group homes).
- Adults may be users of community services.
- Adults may be users of services for people with learning difficulties.

Patterns of communication

- Early symbolic communication needs to relate to their own items/events.
  - The photograph may need to be of *their own* cup, not any cup.
  - The gesture is the one you use together.

  Shaping into more conventional means may be very slow, if achievable at all.
- Deafblind people may be at several stages of communication at one time. They may use one or two signs, understand a few pictures but mostly make themselves understood through tears and smiles. Communication passports are useful to describe this.
- Deafblind people may communicate more fluently with some people more than others, and be reluctant to communicate at all with some individuals.

A competent communication partner

To be a competent communication partner, who enhances the communication skills of a congenitally deafblind person, you need to:

- Give plenty of time – allow them to respond at their own pace. Deafblind people have lots of patience.
- Have plenty of different communication methods available and be prepared to swap between them.
- Try to express something in a less symbolic way if needed.
- Try to pair communication in more and less symbolic ways.

- Look for meaning in all situations – try to interpret even the earliest communication behaviours and act on them.
- Allow the other person to make contributions, even if you don't understand them.
- Have fun – be an interesting person.
- Have high expectations.
- Be prepared to follow the other person's lead and respond to their communication. (Why shouldn't they have a shower at 3pm if they want to? Just because you like to have one in the morning, doesn't mean they do.)
- Ensure you both make the best of their residual vision and hearing.
- Don't make them dependent on your prompting (asking questions, touching a hand) to communicate. Be ready to be a communication receiver – *they* may be communicating to *you.*
- Ensure resources are available as required – symbol books, voice output aids.
- Ensure their vision and hearing needs (quiet environment, light from behind, etc) are taken care of.
- Use any aids which are helpful – sound field system, hearing aids, glasses, CCTV.
- Think about new vocabulary they might need.
- Ensure that they have experience of you and other competent communicators using the methods they use to communicate, so they can imitate. Share the communication, give it back.
- Ensure that they have the opportunity to experience 'vocabulary' before they are expected to produce it.