

# CHARGE Syndrome

## Educational, behavioral, & developmental aspects

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# What is special about CHARGE?

- Strong common characteristics BUT a very wide range of ability and function
- Large number of anomalies (up to 38?)
- True multi-sensory impairment
- Frequently high developmental potential plus features that normally suggest low developmental potential
- Brilliant adaptive abilities but also bewilderingly patchy development
- Sensory integration dysfunction that affects all areas of development, learning, and behavior
- Significant difficulties with self-regulation and with executive function
- Inherent high levels of stress

David Brown - Am.J.Med.Gen. 2005

“Children with CHARGE syndrome are truly “multi-sensory impaired”, having difficulties not only with vision and hearing but also with the senses that perceive balance, touch, temperature, pain, pressure, and smell, as well as problems with breathing and swallowing, eating and drinking, digestion, and temperature control.”

David Brown - Am.J.Med.Gen. 2005

“...the immense difficulties that children with CHARGE face in almost everything that they do, and, as a consequence, ... the very high levels of stress with which they must live for much or even all of their lives. Time spent trying to reduce stress levels, and trying to give the children acceptable strategies for doing this for themselves, must be one of the most precious gifts we can offer them, and one of the biggest favors we can do ourselves as educators, therapists, and family members.”

# 1981 - 6 anomalies

- C - Coloboma
- H - Heart Defects
- A - Atresia of the Choanae
- R - Retarded Growth/Development
- G - Genito-urinary Defects
- E - Ear anomalies

## 2006 - 38 anomalies???

- Mutation of gene CHD 7
- Facial Palsy
- Cranial Nerve anomalies
- Semi-circular canal anomalies
- Dental anomalies
- Sense of smell anomalies
- Larynx & Pharynx anomalies

## 2006 (Continued)

- Atresia of the esophagus
- Skeletal anomalies
- Sleep apnea
- Tracheo-esophageal fistula
- Cleft lip / Cleft palate
- Hypocalcaemia
- Seizure disorder

# Cranial Nerves

- I Olfactory
- II Optic
- III Oculomotor
- IV Trochlear
- V Trigeminal
- VI Abducens
- VII Facial
- VIII Vestibulocochlear
- IX Glossopharyngeal
- X Vagus
- XI Accessory
- XII Hypoglossal





# Cranial Nerve Anomalies

- Nerve 1 - Smell (42%)
- Nerve 2 - Vision (80%)
- Nerve 7 - The Face (43%)
- Nerve 8 - Hearing & Balance (80%)
- Nerves 9 and 10 - Swallowing (50%)

(From CHARGE Foundation Parent Manual - 1999)

# CHARGE - the most 'multi sensory impaired' of all syndromes

Problems with the perception of:

- Vision
- Hearing
- Touch
- Proprioception
- Temperature
- Pain
- Vestibular
- Smell
- Taste

## David Brown on CHARGE - 1997

“I know of no identified sub-group within the population of people with multi sensory impairment who have so many medical problems, of such complexity and severity, and with so many hidden or delayed difficulties, and yet no sub-group has shown such a consistent ability to rise triumphantly above these problems”







06.10.2006

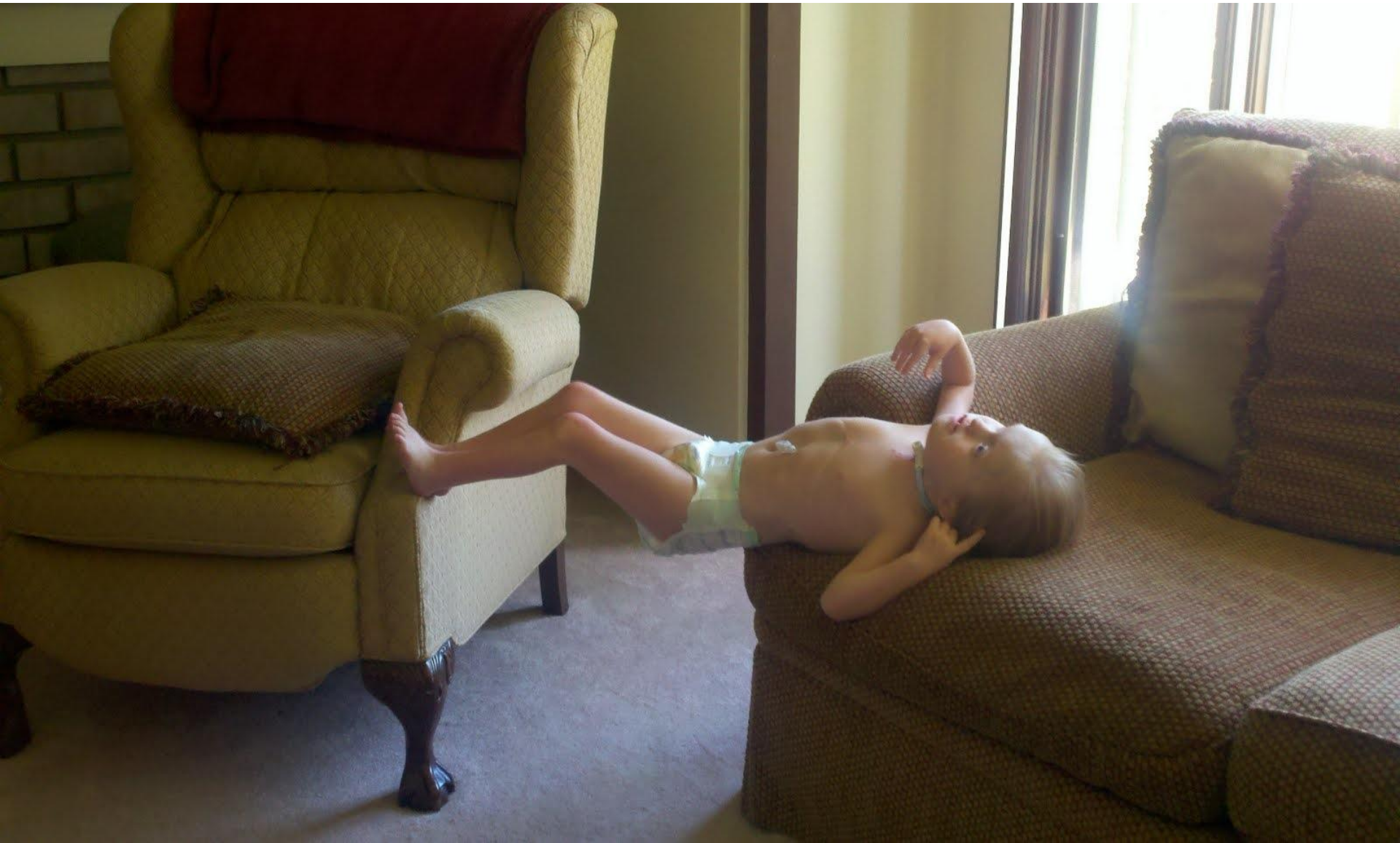


















This is normal viewing posture...

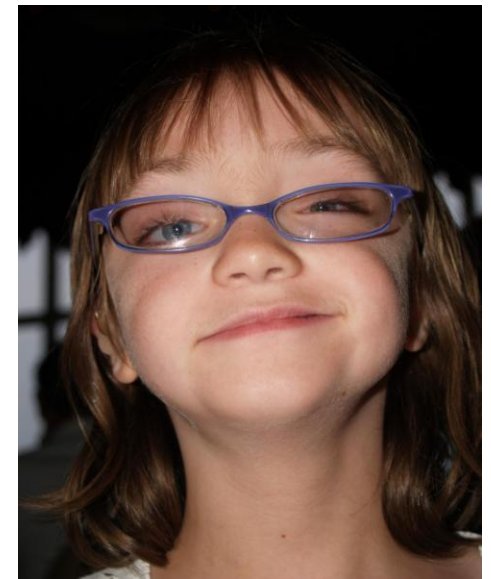
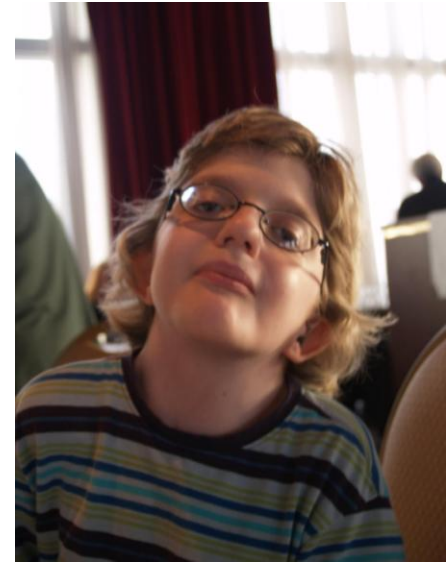
...when you have no vestibular sense, upper visual field loss, poor tactile & proprioceptive perception, & low muscle tone.





# Mental retardation? Or....

- No balance sense
- Low muscle tone
- Visual impairment
- Hearing impairment
- Facial palsy
- Executive function deficit





# Mental retardation? Or....



- Breathing difficulties
- Eating difficulties
- Illness, hospitalization
- Sensory defensiveness
- Inappropriate assessment

# Obstacles to the clear articulation of speech for people with CHARGE (1)



- Hearing Impairment
- Vision Impairment
- Facial Palsy
- Low muscle tone
- Poor tactile sense
- Oro-facial clefting
- Enlarged Tongue
- Poor tongue movement

## Obstacles to the clear articulation of speech for people with CHARGE (2)

- Small lower jaw
- Larynx/Pharynx anomalies
- Breathing difficulties
- Swallowing difficulties
- Dental anomalies
- Delayed/immature eating skills

## Obstacles to the clear articulation of manual signs for people with CHARGE (1)

- Low or poorly modulated muscle tone
- Poor tactile sense
- Poor proprioceptive sense
- Poor spatial awareness
- Dyspraxia
- Poor body awareness





## Obstacles to the clear articulation of manual signs for people with CHARGE (2)



- Poor bilateral coordination
- Poor sense of balance
- Various postural difficulties
- Low vision or blindness / visual field losses
- Skeletal anomalies





# Educating children with CHARGE syndrome in local schools

1. Resources, literature, advocates
2. Deaf-blind education
3. CHARGE-specific considerations
  - Health issues
  - Self-regulation and executive function issues
  - Language and communication issues
  - The role of the Intervener

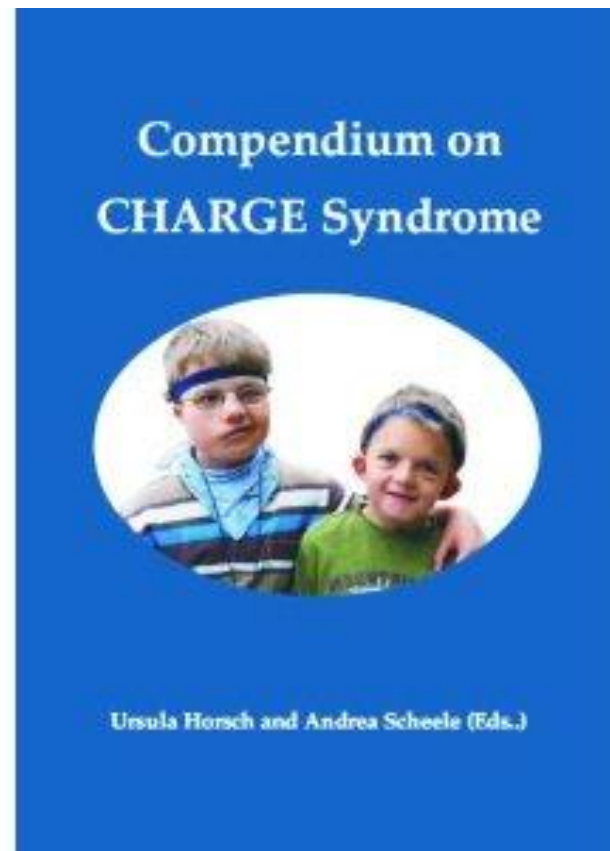
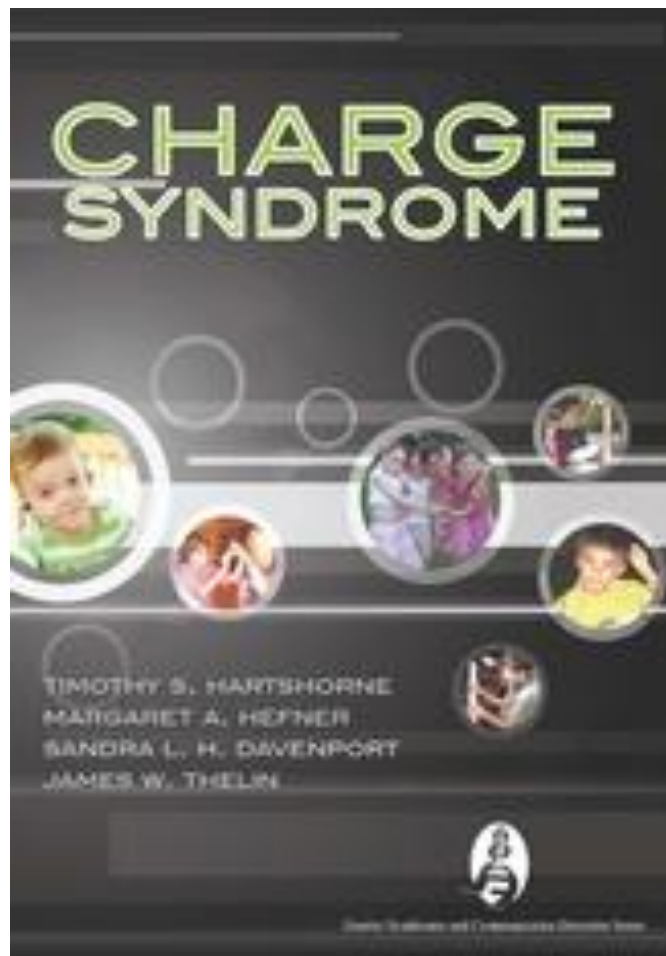
# Individualization

“I would argue that what people with an intellectual disability need more than anything else is to be accepted and respected ***as they are***. The aim of all of us who engage with them should be to support ***who they are***, to provide the supports so they can be ***who they are***, and to interact with them in such a way that ***their ways of being*** are appreciated and nurtured rather than undermined and dismissed. What this requires is stretching our rules of engagement and intimacy.”

Jani Klotz

# Resources, literature, advocates

- CHARGE Syndrome Foundation (Professional Packet, AJMG, ASHA video, Parent Manual, CHARGE Accounts, Links)
- Perkins School for the Blind CHARGE webinars
- CHARGE Lab at Central Michigan University
- DB-LINK
- Texas School for the Blind
- California Deaf-Blind Services
- Books from the USA and from Germany



# Deaf-blind education

## Educational Needs of Children with CHARGE Syndrome

Martha M. Majors, M.Ed. Sharon Stelzer, M.Ed.

<http://chargesyndrome.org/professional%20packet/11%20educational%20needs.pdf>

Martha Majors & Sharon Stelzer  
Educational Needs of Children with CHARGE  
Syndrome

Communication

Impact of sensory losses

Curriculum

Environments

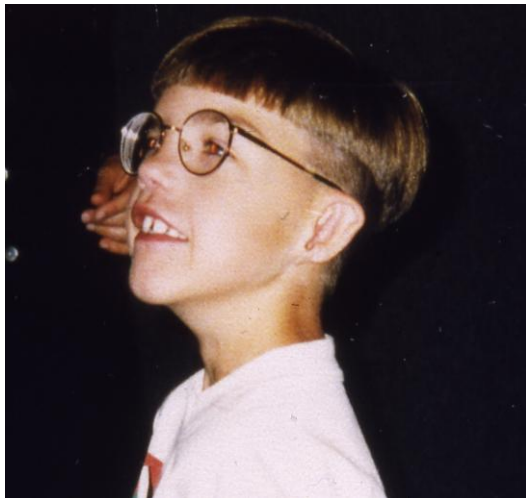
Teaching strategies



# What might be needed? (1)

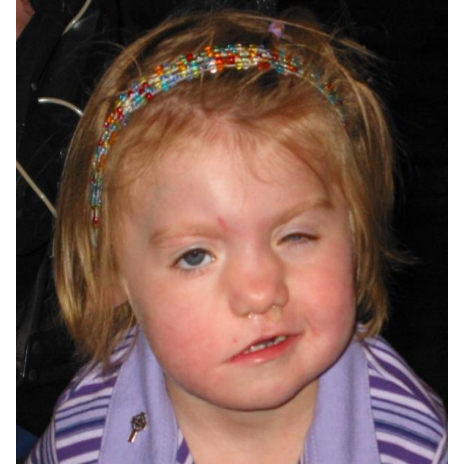


- 1-on-1 support
- Sensory Impairment Team (Services for Blind, Deaf, Deafblind, as appropriate)
- Physical Therapy, Occupational Therapy, & Speech Therapy support
- Sensory Integration program
- Adapted furniture
- Controlled or adapted environments
- Consistent routines
- Functional activities



## What might be needed? (2)

- Individualized motivators
- Appropriate communication systems (possibly multi-modal, including a concrete system)
- Individualized pacing
- Facilities for safe rest periods
- Specific support for group sessions
- Nursing or paramedical support
- A teacher who has 'IT'



# Health issues

Complex health issues

Availability of medical/para-medical services

Balancing health and educational needs

Making health care educational

# What is CHARGE Behavior??

- Impulsivity
- Obsessions
- Self Stimulation
- Poor self-regulation
- Executive function disorder
- Attention Deficit Hyperactivity Disorder
- Pervasive Developmental Disorder
- Obsessive Compulsive Disorder
- “Autistic-like” behaviors
- Deafblindness
- Multi Sensory Problems.....(and so on!)
- Laziness, stubbornness, aggression





## Self-regulation/executive function issues

[Self-regulation]... “is defined as the capacity to manage one’s thoughts, feelings and actions in adaptive and flexible ways across a range of contexts”

Jude Nicholas, CHARGE Accounts, Summer 2007

# The 9 levels of arousal

(Carolina Record of Individual Behavior)

- Uncontrollable agitation
- Mild agitation
- Fussy awake
- Active awake
- Quiet awake
- Drowsy
- Active sleep
- Quiet sleep
- Deep sleep



# Self-Regulation

Can we help the child to recognize and deal with excessive levels of over-arousal or under-arousal, in socially acceptable ways?

If self-regulation is difficult, can the child learn ways of asking for help?

Does poor sensory perception (ie. poor or delayed or fragmented feedback) explain...?

- Need/preference for strong inputs
- Apparent sudden and extreme reactions to pain (I had no idea I was hurting until I was in agony)
- Apparent sudden and explosive changes in emotional state (I had no idea I was angry until I was furious/ I had no idea I was scared until I was terrified)
- Apparent executive function problems
- And is impulsivity an outcome of the above - or sometimes a functional solution?



## David Brown - Am.J.Med.Gen. 2005

“Many people with CHARGE demonstrate difficulties with vocabulary recall, initiating communicative exchanges, and with clearly articulated expression, in the abstract forms of spoken and/or sign language. Provision of a communication mode with a concrete component (eg. objects, symbols, pictures, written words) can be of immense help in aiding recall, in encouraging initiations, in clarifying meaning, and in generally fostering a more confident, animated, and fluent communicative style.”



## “Exploring executive functioning”-Amanda Kirby

- **Activation** – organizing & prioritizing, initiating, getting started
- **Focus** – sustaining & shifting, completing
- **Effort** – regulating alertness
- **Emotion** – managing frustrations, modulating emotions, keeping perspective
- **Memory** – remembering, accessing recall, recognizing & remembering a sequence
- **Action** – monitoring & regulating self-action without impulsivity, or poor context or poor pacing

# Language and communication issues

Multi-modal

Include concrete modes

Other supports (eg. calendar, word cards, vocabulary book)

“The child’s preferred modes”

Mixed and/or varying modes

# The role of the Intervener

NEED: SOCIAL & EMOTIONAL WELL-BEING

*Role of the intervener:* to develop and maintain a trusting, interactive relationship that promotes social and emotional well-being.

In my work with children with CHARGE I am always thinking about their “*sensory comfort*” and their “*sensory confidence*” .



# Comfortable Shoes



# Ways & Means Case Study

# The student

- 8 years old girl
- Multiple health issues with many early surgeries & hospitalizations
- Suppressed immune system - frequent infections
- Severe balance issues, with delayed age of independent walking
- Fed completely by G-tube
- Some hearing – she has some speech perception with amplification
- Lip reader
- Primarily an American Sign Language user
- Visual acuities 20/400 in both eyes + visual field loss

# School

- Program for deaf students with additional disabilities
- The Director of the service was flexible, imaginative, persistent, and supportive of students and staff
- Large support team (e.g. Deaf Teacher, Behaviour Specialist, Physiotherapist, Occupational Therapist, Speech Therapist, Adaptive Physical Education Teacher, Nurse, Psychologist, Sign Language Interpreter, Social Worker)
- High ratio of adults to students in the classroom
- Teachers & aides were all very highly qualified and experienced



# School (continued)

- Strong emphasis on communication and language
- Total Communication approach, plus some ideas from the literature on CHARGE (eg. daily schedule of symbolic objects & pictures)
- Student had her own aide due to behavioral issues
- High level of academic expectation in the class
- Student spending increasing proportion of the day in individual activity with her aide because group sessions were very challenging for her
- Challenging behaviors were increasing in severity & frequency

# The issues

- Often ***impulsive*** in behavior - e.g. escaping and running away, sweeping tables clear of objects, throwing objects, physical attacks on adults and (especially) other students
- All the above behaviors could also be used with intention in a planned way
- Alternation between extreme ***passive compliance*** and extreme ***non-compliance*** - described as “wanting to please people and wanting to upset and hurt the same people”
- Clearly an ***intelligent*** student yet also with perceived ***cognitive issues*** - e.g. unable to attend for long, unable to retain or generalize concepts learned, huge ***variability in skill level*** (from moment to moment and from day to day)
- Strong dependence on familiar, ***predictable routines***, with behavioral outbursts if her expectations were not met
- ***Ritualistic behavior*** in familiar activities
- Frequent insistence on ***getting horizontal*** on her back on the floor and being left alone for short periods of time

## Before my visit - what had worked?

- Providing a '*safe area*' next to the window for her 'horizontal' periods
- Increase in the number of Adapted Physical Education sessions per week because of the beneficial after-effects of these sessions
- Daily calendar of objects & pictures
- Sessions with her favorite aide in the classroom (who was not her regular 1-on-1 aide)
- Increasing the time spent in individual sessions with less time working in a group
- Being taken for a walk around campus after more severe behavioral events or outbursts

## Before my visit - what had not worked?

- Physical restraint in a chair
- Being taken for a walk around campus after her more severe behavioral events or outbursts
- Extending the duration of a lesson to complete the task if it had been disrupted by her behavior
- Limiting the availability of time with her favorite aide to reduce the risk of her developing 'over-dependence'
- Limiting & scheduling the availability of the 'safe area' next to the window for 'horizontal' periods
- Behavior plans (from 2 different specialists) that depended primarily on the use of punishers & on a token system
- Intensifying homework to facilitate better academic achievement & attention in class



## My perception of what was wrong (1)

- There was no recognition that this is a student with deafblindness
- There was too much pressure to '*perform*' like her peers who are 'only' deaf, because she is a clever girl with very good adaptive skills, so her challenges were easy to miss or easy to under-estimate
- Failure to consider her individual motivators, and then use them within appropriate areas of the curriculum
- Failure to recognize her extended processing time and processing strategies (e.g. standing up to pay attention and learn, walking around the room briefly to process)
- Insistence that the student sit upright on a regular chair at a regular table for working on many areas of the curriculum

## My perception of what was wrong (2)

- Not understanding the need for her to move and vary her position, and to perform tasks that provide strong proprioceptive input, to assist with her poor attention and poor self-regulation (& postural insecurities)
- Expecting the student to work at the thresholds of her sensory abilities most of the time
- The deliberate involvement of a variety of adults to prevent over-dependence on one adult in the school setting
- Team anxiety about using the “*follow the child*” idea in case this would spoil her, or increase her non-compliance
- Persistent use of behaviorist methods to control non-compliance and impulsivity, using a succession of practitioners, none of whom had familiarity with CHARGE nor with deafblindness, “....which just made the problems much worse”

## What improved the situation? (1)

- The label of '***Deafblind***' - providing access to more appropriate expectations, assessment approaches, teaching strategies, and resources
- Provision of an Intervener (who was already her favorite adult in the school)
- The use of the student's individualized motivators in as many curriculum areas as possible
- "*Follow the child*" as a guiding philosophy
- More careful '*reading*' of the student by staff, to identify and notice the precursors of stress, tiredness, and over-arousal (and ideas about what to do in response to these problems)

## What improved the situation? (2)

- Involving the student in making a Personal Passport so that other people (adults and peers) from outside the class could '*tune in*' to her quickly and appropriately
- More precise, carefully chosen, & carefully presented, choices so that the student could feel more empowered and in control, & not so threatened and confused
- Provision of information and ideas about the vestibular and proprioceptive senses, and their impact on behavior (especially on attention and arousal), then putting this knowledge to use in both structured and opportunistic ways (involving Adapted Physical Education, Occupational Therapy, and Physiotherapy specialists)



## What improved the situation? (3)

- Regular movement was encouraged, tolerated, and observed for its impact, once the relationship with the Intervener had developed and non-compliance had reduced accordingly
- The existing '*safe place*' for getting horizontal and staring at light was made accessible throughout the day rather than only for short periods at times pre-determined by a fixed daily schedule
- Determined efforts were made to link home & school effectively through daily contact

