

12th CHARGE- Conference

22nd – 24th June 2018

*Youth hostel
Oberwesel,
Germany*

Rob Last

22nd June – 2.30 p.m.

“Friendships, Belonging and Connection”

Over the years in discussions with parents the topic of ‘friendships’ has arisen and the challenges children and young adults sometimes experience in developing and sustaining friendships.

Often, apart from the family, it is the medical, therapy, early intervention and education teams who become the friendship circle rather than kids from the neighborhood.

In this presentation I will explore friendships, belonging and connection.

This will include discussion on diversity and difference, self-esteem and confidence, peer acceptance, bullying, building capability and strategies for nurturing friendships at home and at school.

Friendships, Belonging and Connections

CHARGE Syndrome Conference, Germany

Oberwesel

21 – 24 June 2018

Rob Last AM

Introduction:

In a recent CHARGE syndrome CHARGE Accounts Newsletter USA, there was an article with the title 'A New Friendship' by Molly Roberts.

The family were attending the Orlando conference in 2016, without siblings, just Mum, Dad and Christian.

In the article the words connecting and belonging came up.

Molly wrote:

'Typically Christian prefers to stay on his iPad and not socialize, however this night we encouraged him to at least say 'Hi' and share his name with his peer group. Quickly he met Andrew, another teenager with CHARGE. They had a lot in common – interests in 'How to Train Your Dragon' and 'iPads'. These two boys sat by each other and communicated through sign and their iPads. Their interaction was unprecedented and we decided that creating opportunities for their **connection** to grow would be the focus of our weekend.

Over the weekend the boys kept the **connection** alive'

and

'At the conference we always feel acceptance and immediately feel we **belong**.'

In my experience this is what occurs at all conferences, here in Germany, in Australia and New Zealand and the USA.

Belonging and Connections:

What's the difference?

Belonging relates to the symbolic spaces which feel familiar, comfortable and secure, and to which a person feels emotionally attached. We develop a sense of belonging over time, remembering experiences, people and places.

Connectedness refers to the quality and number of connections with people and place.

Key elements in helping a sense of belonging and connection are friendships, acceptance by peers, feeling valued and capable, having supportive relationships with key adults at home and at school.

This describes these conferences.

In the beginning:

For people who have CHARGE syndrome, belonging and connectedness may develop very differently.

For young people who have CHARGE it is likely that the first friendship circle outside of the family will be the medical, therapy, early intervention and educational teams, rather than the kids from the neighbourhood.

Connectedness may be with family; parents, siblings, grandparents and the medical and therapy team, which includes varying doctors, occupational therapists, physiotherapists, speech therapists, nutritionists, special education teachers and more.

Once out of the medical environment the family remains the significant others and the home visiting therapy team may be seen as other primary connections. As we all know these are not the typical circles of friendships other children may experience.

In time playgroup, preschool/kindergarten and school become the beginnings of new connections and in time a sense of belonging.

These friendships with teachers and therapists can be rich and rewarding and last for many years. For me I continue to be friends with kids I met in the first few months of their life, who are now in their teenage years, twenties and thirties.

Friendships with peers are really important and are to be encouraged and nurtured

Diversity and Difference:

At these conferences, diversity and difference are commonplace with acceptance of difference being the norm.

I'm sure you all relate to the issue of people looking at your child in a different way. In talking with parents they describe the challenges:

‘On good days it’s OK, on not so good days it bothers me’.

Here, we might no longer see the differences in the children and adults, however when we are out and about we may notice the responses of others and once again see the differences.

This probably happens all the time, at preschool, school and when they are out and about, just an everyday occurrence.

Being of different appearance may impact on the connections children make at preschool and school and when they are out in the wider community.

Their peer group and the general public may not be as unseeing.

I notice in our children with CHARGE they don’t seem to particularly notice, they pose for photos confidently, accept to speak at conferences, mix well socially with familiar folk, walk down the street with pride and don’t seem to notice or care what people notice.

However being of different appearance may impact on the connections children make at preschool and school and with the kids in their local community as well as their self-esteem.

An interesting modification I’ve noticed in a young person I know well, she has modified her smile to a grin rather than smile broadly, as her facial palsy was more pronounced in a broad smile.

Photos: Four photos. Sophie and Sean

To expand the concept of difference and diversity, I’ll use the example of an Australian woman who was severely burnt in a grass fire while competing in a 100 kilometre ultramarathon in 2011.

Her name is Turia Pitt. She has become an advocate for people who are of different appearance.

Slide: Turia

She is a superstar of difference and diversity.

Her self-image is intact, self-esteem in place, pride in who she is and is willingly advocating diversity and difference.

All the things we want for our kids.

Last year I saw three movies, which focussed on being of different appearance. Wonder, The Shape of Water, Beauty and the Beast.

Slide: Movies focusing on people of different appearance.

Movies focussing on people of different appearance:

- Wonder - 2017
- The Shape of Water - 2017
- Beauty and the Beast – 2017

Slide: Building and Nurturing Friendships

Building and Nurturing Friendships:

- Frequent, emotionally pleasant interactions need to occur and these interactions need to happen in a relatively stable framework of concern for each other.
- **In the mainstream school environment** where the student is in an inclusive program, the strategies the school has in place may be a buddy system, a mentor program, special interest groups, and support workers in the classroom, who encourage friendships.
- This is not the same as forming a friendship through common interests and the desire to develop a friendship through mutual engagement.
- For the student with CHARGE who doesn't hear all the nuances, can't see the details, misses the subtleties of facial expression and body language; which may result in confusion as to what is going on, not being able to keep up, feeling left out and perhaps giving up and withdrawing.
For them forming and nurturing friendships can be a challenge.

In the special school environment I've observed different outcomes. When students are with other students who have varying special needs it can seem like they have 'found their tribe'.

Slide: Finding your tribe

Finding your tribe:

I attended a seminar on 'Facial Difference' where the speaker had significant facial difference.

She spoke of the importance of "finding your tribe".

At these conferences I see everywhere where the kids seem to have 'found their tribe'. Just like Christian and Andrew, whom I talked about earlier.

Throughout my career as an Early Childhood Educator

I was a strong supporter of inclusive education. In particular in the early years, during the playgroup/kindergarten/preschool years. In these educational environments the kids are surrounded by regular kids with regular behaviour, with regular speech and language, with regular play and regular expectations.

I advocated this for children of all abilities.

It was their chance to be with the local kids, to be part of the community, an opportunity to form local friendships, to be invited to local kids birthdays and also to be immersed in the regular behaviours of kids.

However I've had to rethink this in regard to the whole educational journey. Once embarking into the next stages of education primary and secondary school, the inclusive school program becomes increasingly challenging.

I've observed parents decide to begin with mainstream, and at some stage in the school journey to make the change to a specialist school environment, or to adopt home schooling.

Slide: Sean and Ethan

In the special school environment I observed Sean and Ethan 'finding their tribe'.

Sean followed mainstream education up until year 10, in year 11 he transferred to a special school.

Sean's literacy skills are age appropriate however the academic curriculum and managing the higher school education challenges were too great.

He and his parents made the choice to transfer to a special school. Sean was involved in the process and was able to spend a couple of days a week at the special school to get a feel for it. To ascertain if this was going to be suitable for him. At the special school he became school captain, voted for by his peers, and he found a good friend.

At this school he 'found his tribe'.

Slide: Strategies at school

Strategies at school:

There are strategies to prepare the student and the school, no matter what school it may be.

- Orientation to the school. Several visits prior to commencement. School needs to be a familiar and safe place.
- Orientation to the school staff. They need to be knowledgeable on the challenges of CHARGE syndrome and familiar with the student.
- Involve the student in all the school commencement process.

- Highlight the skills and interests of the student:
communication, academic, personal, social, recreational, leisure, sport, hobbies and community involvement.
- Encourage a buddy system.
- The aim is for the student to:
Feel capable. This applies to academic achievements and non-academic achievements.
Feel valued. This is being appreciated for the qualities that the student brings to individuals and the wider school community.
- Encouraging involvement in common interest groups.
- Finding the supportive students and adults at school.
- Nurture connections when they occur.
- Encourage initiative and independence.
This is trusting that students have the capability to manage their social times with autonomy whenever they choose, this may be with other students or may be by themselves. At other times they may need the support of an adult.
It is usually a mix of both.
- Circle of Friends
Circle of friends is an approach to enhancing the inclusion, in a mainstream setting, of any young person (known as 'the focus child'), who is experiencing difficulties in school because of a disability, personal crisis or because of their challenging behaviour towards others.
inclusive-solutions.com

Strategies at home:

In 2016 I spoke about focussing on 'passions' and then using the 'passion' to find those who have a common 'passion', which may result in developing social connections and friendships through common interests.

- Notice the passion, nurture the passion, and embrace the passion.
- Find individuals, groups and clubs who have the same passion.
- Use the passion as an educational opportunity and as a possible employment opportunity.

Thoughts for parents?

These apply to the transitions from home to educational settings and to changes in educational settings: home to preschool, preschool to primary school, primary school to secondary school, secondary school to tertiary education, secondary school to employment options.

For parents it's about:

- Networking, energy, perseverance, research, positive outlook, assertiveness.
- An ability to advocate the student's strengths, gifts and talents.
- An ability to describe the students needs, challenges and supports required.
- Considering and describing the ideal school/work day for the student.
- A clear understanding of what is not acceptable for the student's future.

More tips for parents:

Youtube video with Zach Anner

Bullying:

- The research literature shows children with disability experience more bullying than peers, regardless of age, educational setting, gender or type of disability.
- Establish the school/workplace policy on bullying.
- Use students who have been bullied to educate other students about bullying prevention.
- Friends provide a 'wall of support' and protection.

Time alone:

- Something I admire in people with CHARGE is their ability to appear comfortable in their own space.
- To spend time alone doing what they like to do.
- To not seek lots of social experiences.
- We may perceive them as feeling lonely but this may not be how they feel.
- Give permission and allow for time alone.

The Internet:

- Researchers are increasingly recognizing the potential of the Internet to provide social and emotional support to various groups that share a common goal, identity or experience.
- The Internet is the most effective tool for children and adults with CHARGE syndrome because they are spread across the globe and frequently not mixing with other children within their immediate community.

- The Internet, therefore, provides a central meeting place for those who are unable to regularly meet with similar others at a single geographical location.
- Benefits of online communities include increased recognition and acceptance from others, reduced social anxiety, reduced isolation and strengthened social support networks, and a greater acceptance of one's own identity.

In conclusion:

This a short story about belonging and connecting told through pictures.

Photos: Cassandra and Sean 'doing lunch'.

- Cassandra and Sean met at a picnic.
- They hadn't met before but connected quickly and easily.
- It was time for lunch.
- Their lunch was prepared.
- Cassandra realized Sean's lunch was just like her lunch.
- They enjoyed this experience very much.
- Sean's arm crept around Cassandra's, connection and belonging was complete.

In addition:

... determination and willpower

David Brown in his famous quote, spoke about iron willpower, this photo depicts it perfectly.

... and charm

... friendships, belonging and connecting

Thank you

Rob Last

Tim Hartshorne

22nd June – 4.15 p.m.

“Five hints for new and seasoned parents”

Parenting is always challenging, but particularly with a child who has challenges, such as one who has CHARGE. This presentation reviews five considerations for parenting a child with CHARGE



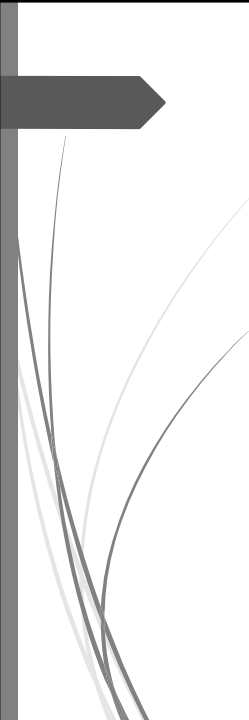
Five Hints for new and seasoned Parents

Raising your child with CHARGE

Timothy S. Hartshorne

Communication, communication, communication

With oneself, with others, with the world



Communicate as you would with any other baby

- Facial expressions are important
- Cooing and laughing
- Motherese
- Touch
- Imagine being in a foreign country and not understanding the language



Learn the communication system that works best for your child

- Not the other way around
- Do not use just one system
- Feel free to combine communication approaches
 - Gestures and facial expressions
 - Oral
 - Sign
 - Picture

“My child has no communication”

- This is never the case.
 - All behavior has a purpose
 - All behavior is communication
- Once the child believes nobody is communicating, they will shut down
- Not all behavior is intentional communication – but it can be read
- Even if your child likes to withdraw, do not let them alone for too long

Routine



Find a routine as soon as possible

- Children with CHARGE thrive on routine and predictability
- A calendar system to communicate the schedule is essential
- While a certain amount of chaos is quite natural, particularly in some families, it will not be compatible for your child with CHARGE

Anxiety is related to uncertainty about what is going to happen next

- What will happen today?
- What do they want me to do?
- Where are we going?
- What will happen when we get there?
- Will I feel **safe**?
- The world does not always make sense
- It can be unpredictable



We like to know what is going on.

- What are we doing right now?
- What are we going to do next?
- What did we just do?



When we are not sure...

- We become anxious
- We engage in behavior which expresses our feelings
- We engage in behavior or activities that help us feel more secure

Hitting
mom



shutterstock · 211012960

Supporting self-regulation

- Self-regulation allows us to organize and manage our thoughts, feelings, behavior, and body, so that we can manage our day and meet our goals
- Because self-regulation skills are hard for children with significant disabilities to develop
- We provide the external support for what will become an internal self-regulatory process
- A major component of this is routine



Discipline

"I can't say 'NO'."

No pass on discipline

- Because your child has CHARGE does not give them a break on learning to cooperate with family living.
- Behavior is communication

Jacob and the bath





Behavior also has consequences

- Natural consequences – if I did nothing what would happen
 - Doing nothing is surprisingly hard work for parents
 - Don't want to appear to be a bad parent
 - It can mean extra work
 - If you push your food off your plate, it falls to the floor and the dog eats it.
- Logical consequences – pre-established response
 - "Let the punishment fit the crime." Teaching cause and effect.
 - Must be related to the child's action
 - Must be revealed in advance
 - The child chooses the consequence
 - Splash in the tub and bath time is over



Destructive and Aggressive Behavior

- Consequences do not work very well
- First, stay calm
 - Of course it is not easy
 - But if you get aroused, it will increase the arousal of the child
- Know your child and their triggers
 - Changes in routine
 - Chaotic environment
 - Pain or illness
- Can the environment be modified to minimize risk?
- Watch out for responses to the child that increase the likelihood they will do this again



Getting Connected

Who understands what you are experiencing?



Get connected

- It is hard enough to raise a more typical child. For this one you need help.
- Join the CHARGE Syndrome Foundation
- Get together with other families who have a child with CHARGE
- Go to the conferences
- Join the Facebook pages
- Connect with parents of children with other disabilities
- Check out services for children who are deafblind



Acceptance and Advocacy

Acceptance and Advocacy

- Your child has CHARGE. It is tough on everyone. Move on.
- Learn to accept and love your child with no need for your child to be any different
- You are your child's primary and often sole advocate
- Your job is to fight like heck to make sure your child gets what he or she needs
- Find allies in the fight

TAMING THE WOLVES

AND THE PARENT FROM HELL



Diagnosis



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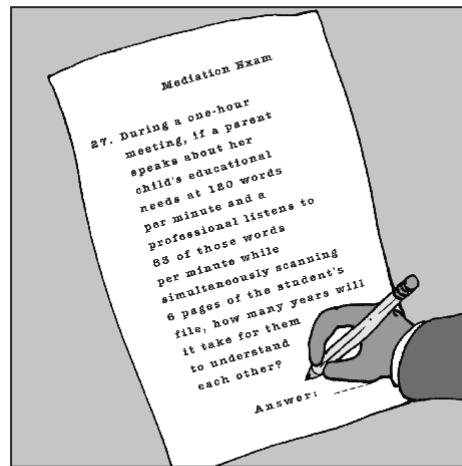
Treatment



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THE DISTRICT'S EXPERIMENTAL METHOD
FOR DETERMINING RELATED SERVICES
WORKS JUST AS WELL AS MAKING THOSE
DECISIONS BEFORE KNOWING A
STUDENT'S IEP GOALS AND OTHER
LEARNING OUTCOMES.

Communication



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WORD PROBLEMS

Intimidation



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OUTNUMBERED?

Predictions



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DECISIONS ABOUT A CHILD'S LIFE:
SOMEONE IN THE FAMILY SHOULD BE
WEARING THE PANTS.

MARATHON SKILLS

Ann P. Turnbull



- Meet basic needs
- Know your self and your family
- Love unconditionally
- Establish relationships
- Experience and benefit from emotions
- Take charge
- Anticipate the future
- Establish balance

David Brown Tim Hartshorne

23rd June – 2.00 p.m.

***“CHARGE Syndrome and Behavior:
Diagnosis and Intervention (Part I)”***

Diagnosis of behavior. Individuals with CHARGE are often diagnosed with a variety of psychiatric disorders such as autism, OCD, and ADHD. This session addresses the problem of psychiatric diagnoses in children with CHARGE: what are these diagnoses, why are they given to children with CHARGE, and what problems do they create, and are they appropriate?

CHARGE Syndrome and Autism, ADHD, ODD, OCD, Et al.

*Tim Hartshorne, David Brown, and
Shanti Brown*

That's weird; must be autistic



Diagnoses Piled On

- Autism
- ADHD
- OCD
- Tourette
- Etc.



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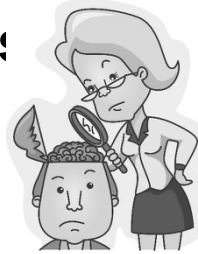
Problems with these Diagnoses

- Create a reality that may not exist
- Lack explanatory value
- Ignore the uniqueness of the behavior
- Lead to multiple drug treatments



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What are mental disorder:

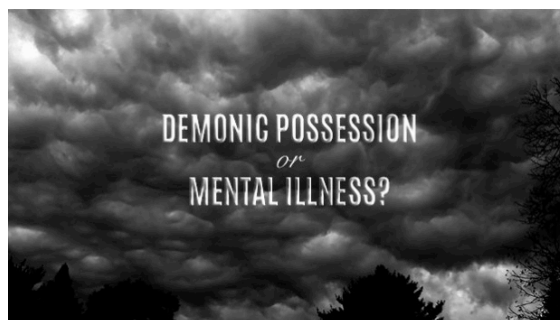


- Nobody really knows
- There are no blood tests
- Symptom co-variation
 - If you have these behaviors you are likely to have this disorder
- There is no way to confirm
 - Except the agreement of professionals

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What is the cause of mental disorders?

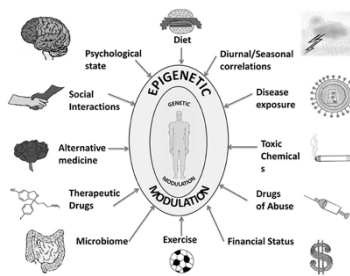
- Probably not evil spirits
- Genetic
- Biological
- Psychological
- Environmental



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Epigenetics – the new direction

- "Certain genes are turned on or turned off, expressed or not expressed, depending on environmental inputs," Richard McNally, PhD, a clinical psychologist at Harvard University



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Diagnosis

- Behavior check lists
- If it looks like a duck, and quacks like a duck



Autism Behavior Checklist

From King, Asch, and Almond (1979)

Patient: _____ Birth Date: _____

Examiner: _____ Test Date: _____

Circle the number for these items most accurately describing the child.

1	Which will for long periods of time	4	3
2	I cannot keep my hand "stagnant" quickly	2	1
3	Frequently does not attend to social/environmental cues	4	1
4	Does not follow simple commands (sit down, come here, stand up, give me)	2	1
5	Does not use toys appropriately (spins wheels, etc.)	2	1
6	Poor use of visual discrimination when learning (takes on parts of objects such as size, color, position...)	2	1
7	Lacks a social smile (may smile out of context)	2	1
8	Exhibits persistent refusal (eyes far from)	3	1
9	Insists on keeping certain objects with him/her	3	1
10	Seems not to hear (ignores verbal hearing tests)	3	1
11	Speech is stilted and awkward	4	1
12	Reacts with the long periods of time	4	1
13	Does not (or did not as a baby) reach out when reached for	2	1
14	Strong reactions to minor changes in routine/environment	2	1
15	Does not respond to own name when called out among two or more other names	4	1
16	Laughs and chatters about, interrupted by crying, no walking, hand flapping...	4	1
17	Not responsive to other people's facial expressions or feelings	3	1
18	Seeks eye "eye" or "if"	2	1
19	Has special abilities in one area - seems to rule out mental retardation	1	4
20	Does not follow simple prepositional commands (e.g., "put the ball in the box")	1	4
21	Sometimes shows no "startle response" to a loud noise	3	1
22	Plays head in or other self-stimulating behavior	4	1
23	Severe temper tantrums and/or frequent minor tantrums	4	1
24	Exhibits aversive eye contact	4	1
25	Reacts being touched or held	4	1
26	Sometimes, painful stimuli (cuts, scratches, bruises) evoke no reaction	3	1
27	Is fixated on a baby, doll, and head to head	1	4
28	Is fixated (above) things when held in arms	2	1
29	Gets distressed objects by examining	2	1
30	Walks on toes	2	1
31	Humms when he brings, kicking...	2	1
32	Repeats phrases over and over again	3	1
33	Does not imitate other children's play	3	1
34	Often will not blink when a bright light is directed toward eyes	1	4
35	Humms with the hand, keeping head...	2	1
36	Does not wait for needs to be met (wants things immediately)	2	1
37	Cannot resist to move than fixated objects	1	4

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Diagnosis



DIAGNOSIS DU JOUR

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Why do we diagnose mental illness?

- As a guide to treatment



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Can people with CHARGE have mental illness?

- Yes
- So why not just diagnose it?
 - Does it lead to better treatment or outcomes?
 - And is the diagnosis accurate?
- Would we expect that the same behaviors which are co-variant in mental disorders for people **without** CHARGE to be co-variant in people **with** CHARGE?
- In other words, what would a disorder such as autism look like in a person with CHARGE?
- Or if a child with CHARGE is said to have autistic-like behaviors, does that mean they have autism?

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Autism

- Impairments in social communication
 - Social reciprocity
 - Nonverbal communication deficits
 - Lack of understanding of relationships
- Restricted, repetitive patterns of behavior, interests, or activities
 - Stereotyped or repetitive motor movements
 - Insistence on sameness
 - Fixated interests
 - Reactivity to sensory input

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Is it autism?

High rates of ASD have been reported in syndromes such as Cornelia de Lange and Fragile X. However, there is debate about whether the ASD profile of behaviours that triggers a Diagnosis in these syndromes is the same as in individuals with idiopathic ASD.

Waite, J., Heald, M., Wilde, L. Woodcock, K., Welham, A., Adams, D., & Oliver, C. (2014).

The importance of understanding the behavioural phenotypes of genetic syndromes associated with intellectual disability. *Peediatrics and Child Health*, 24, 468-472.

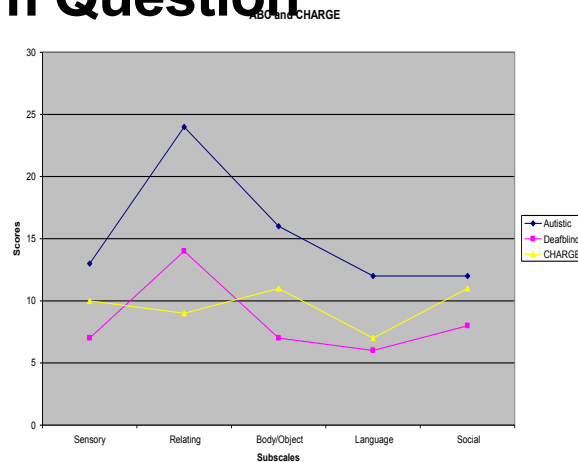
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Syndromes most often associated with autism

- Fragile X
 - social anxiety, extreme shyness and eye gaze avoidance – not central to autism
 - strong willingness to engage socially with others
- Cornelia de Lange
 - The nature of repetitive behaviors appears to be different compared to those with ASD
- Tuberous Sclerosis Complex
 - Repetitive behaviors are not as frequent as in ASD

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The Autism Question

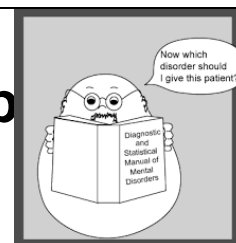


Hartshorne, T. S., Grialou, T. L., & Parker, K. R. (2005). Autistic-Like Behavior in CHARGE Syndrome. *American Journal of Medical Genetics*, 133A, 257-261.

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Autism as an Additional Label

- An array of “symptoms” are seen in CHARGE and Autism, but they differ in function
 - Social: Uninterested vs. not able (Deafblind)
 - Physical: self-stim. vs. regulation
 - Sensory: over-stimulated vs. under-stimulated
 - Communication: immature/uninterested vs. functional
- Autism adds extra label, stops caregivers from finding function for communication



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Why Add the Label?

- Gives parents feeling that something can be done
- “...our goal has been to identify what is unique about the behavior of children with different genetic disorders, and not what is common. I have no doubt that there is a behavioral phenotype associated with CHARGE. My goal, and that of others, is to better specify that phenotype so that we can develop interventions as well as prevention strategies” –T. Hartshorne
- Does an extra label help? How do we treat them differently?

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Gain Access to Services?



- Add label to get ABA services
- Not always a bad thing – but the child with CHARGE could be misunderstood
- More physiological conditions with CHARGE that contribute to psychological behaviors
- ASD has fewer physical abnormalities
- Clinician needs to understand unique physical needs associated with CHARGE versus reluctance often associated with ASD
- ABA services good for FCT but not for everything
 - Walking on grass: irrational fear or painful experience?

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Obsessive Compulsive Disorder



- DSM-5 Diagnostic Criteria for Obsessive-Compulsive Disorder (300.3)

Obsessions

Recurrent and persistent thoughts, urges, or impulses that are intrusive and unwanted, and cause marked anxiety or distress. (Typically irrational fears.)

The individual attempts to ignore or suppress such thoughts, urges, or images, or to neutralize them with some other thought or action (i.e., by performing a compulsion).

Compulsions

Repetitive behaviors (e.g., hand washing, ordering, checking) or mental acts (e.g., praying, counting, repeating words silently) that the individual feels driven to perform in response to an obsession or according to rules that must be applied rigidly.

The behaviors or mental acts are aimed at preventing or reducing anxiety or distress, however, these compulsions are not connected in a realistic way with what they are designed to neutralize or prevent.



We believe.....

- Most individuals with CHARGE syndrome don't have *true* OCD.
 - Someone with true OCD has irrational thoughts leading to irrational anxiety.
 - The treatment involves exposure to the irrational thought, and prevention of the compulsive response.
-
- Individuals with CHARGE have TRUE circumstances that lead to UNDERSTANDABLE anxiety!
 - In other words, the EXPOSURE is happening all the time, and is unavoidable. The compulsive response is an understandable defense for dealing with the anxiety.



OCD-like behaviors seen in CHARGE

- **“Everything must be just right.”**
 - Ordering/lining up/making symmetrical
 - Placing/moving to correct place
 - Light switches
 - Doors closed/open
 - Handing cup to caregivers when he sees it
- **Other things reported or seen:**
 - Repetitive question-asking: “What color is your car?” “What color is your house?”
 - Stuffing things into slots—especially into places from which they are difficult to retrieve.
 - Repetitive, idiosyncratic behaviors: Hand movements, tics
 - Rigid inability to switch activities
 - All-consuming focus on one idea, activity, or item, to the point that it’s not just a hobby/intense interest—keeping in mind that we ALL have hobbies/interests!
 - What else have you observed?

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“A lot of what looks like OCD in CHARGE is really just a reaction to having multi-sensory impairments”— D. Brown 2015

These are actually very creative responses to abnormal, anxiety-provoking circumstances.

Caveat: Someone with CHARGE syndrome could possibly also develop “actual” OCD, but this is not the norm.

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Attention-Deficit/Hyperactivity Disorder

- A persistent pattern of inattention and/or hyperactivity-impulsivity that interferes with functioning or development
- Higher incidence among deaf children
 - Sensory integration issue?
 - Limited language exposure?
 - Social-emotional development issue?
 - Visual selective attention?
 - Boredom?
- Consider executive function impacted by sensory impairments



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Attention Deficit/Hyperactivity Disorder

- | | |
|--|---|
| <ul style="list-style-type: none"> ▪ Diagnostic Criteria:
Inattentive presentation <ul style="list-style-type: none"> • Lacks attention to detail • Lacks sustained attention • Does not seem to listen • Does not follow through on instructions and fails to finish work • Difficulty organizing tasks • Avoids tasks with sustained mental effort • Loses things • Easily distracted • Forgetful | <ul style="list-style-type: none"> ▪ Diagnostic Criteria:
Hyperactivity/Impulsivity presentation <ul style="list-style-type: none"> • Fidgets • Often out of seat • Runs or climbs in situations where inappropriate • Unable to play quietly • Often “on the go” • Talks excessively • Blurts out answers • Difficulty waiting turn • Often interrupts others |
|--|---|

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Attention Deficit/Hyperactivity Disorder

- How can these behaviors be better explained by CHARGE syndrome?
 - Deafblindness
 - Difficulties with executive functioning
 - Attending
 - Organizing activities
 - Inhibition
 - Difficulties with self-regulation
 - Sensory needs

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Attention Deficit/Hyperactivity Disorder

- Example: Bradley, a third grade student with CHARGE syndrome, never seems to be paying attention during math. He spends math class staring up at the lights or inspecting his fingers close to his face.
- ADHD Perspective:
 - Bradley is unable to attend to what his teacher is saying. Bradley is also very fidgety with his fingers.
- CHARGE Perspective:
 - Bradley cannot see the board where his teacher is writing math equations.
 - Bradley cannot hear what the teacher is saying. He also cannot hear what questions his peers are asking.
 - When Bradley flicks his fingers in front of his face, he is getting sensory stimulation from the visual input.
 - Bradley has a hard time transitioning between activities and needs more time to get ready for math.

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Borderline Personality Disorder

- A pattern of instability in interpersonal relationships, self-image, and affects, and marked impulsivity
 - Fears of abandonment
 - Extremes of idealization and devaluation of others
 - Unstable self-image
 - Impulsivity
 - Recurrent suicidal behavior
 - Highly reactive emotions
 - Chronic feelings of emptiness
 - Anger difficulties
 - Stress related paranoia
- May be associated with the experience of severe abuse, neglect, parental conflict

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Tic disorders

- Two types of tics
 - Verbal
 - Motor
- Tourette's Disorder has both
- Worsened by anxiety, excitement, and exhaustion
- About 25% of children will have transient tics
- "Hearing parents and mental health clinicians unfamiliar with typical behaviors of deaf children may have difficulties differentiating the clinical presentation of symptoms of TD from the effects of deafness, as well as in implementing appropriate interventions." (Chovaz, 2013)
- Homemade utterances and gestures may be the result of
 - Language delays
 - Intentional personal entertainment



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Oppositional Defiant Disorder

- What are the diagnostic criteria?
 - Angry/irritable mood
 - Argumentative/Defiant Behavior
 - Vindictiveness

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Oppositional Defiant Disorder

- How can these behaviors be better explained by CHARGE?
 - Communication deficits
 - Sensory difficulties
 - Deafblindness
 - Executive Functioning Difficulties
 - Fatigue

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Opposition Defiant Disorder

- Example:
 - Julie was eating pureed peas for dinner, when all of a sudden, she scooted back her chair and threw the bowl of peas on the ground.
 - ODD Perspective:
 - Julie threw her peas on the ground to defy her caregiver.
 - CHARGE Perspective:
 - Julie was having some gas pain and needed to alert her caregiver that it hurt her stomach to eat.
 - Julie does not like peas and does not want to eat them.
 - Julie cannot stand the texture of pureed peas.

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Bipolar Disorder

People having a manic episode may:

- Feel very “up,” “high,” or elated
- Have a lot of energy
- Have increased activity levels
- Feel “jumpy” or “wired”
- Have trouble sleeping
- Become more active than usual
- Talk really fast about a lot of different things
- Be agitated, irritable, or “touchy”
- Feel like their thoughts are going very fast
- Think they can do a lot of things at once
- Do risky things, like spend a lot of money or have reckless sex

People having a depressive episode may:

- Feel very sad, down, empty, or hopeless
- Have very little energy
- Have decreased activity levels
- Have trouble sleeping, they may sleep too little or too much
- Feel like they can’t enjoy anything
- Feel worried and empty
- Have trouble concentrating
- Forget things a lot
- Eat too much or too little
- Feel tired or “slowed down”
- Think about death or suicide

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Dunn Conceptual Model

Arousal of thoughts, behavior, feelings, sensations	PASSIVE Self-regulation Strategies	ACTIVE Self-regulation Strategies
Habituation	Non-reactive Tune it out	Sensation Seeking
Sensitization	Reactive to Stimuli	Sensation Avoiding

Psychotropic Medications

- Have generally not been researched with children
- No idea about impact on sensory systems
- Have numerous side effects – impact on brain?
- Are often prescribed off label
- Use is not often monitored closely with observation
- Side effects may lead to additional prescriptions
- There is a tendency to add drugs, rather than replace
- Goal should be to be weaned off of them
- May take away a form of communication
- Sometimes they are survival for the family



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The Trouble with Multiple Diagnoses

- Children's functional behaviors may present as symptoms of a mental health condition; however, given the child's multi-sensory impairment, the behaviors are often reasonable choices by the child.
- By classifying these behaviors as a mental health disorder, we may be dismissing the creative strategy the child has come up with to survive within his/her environment.
- Therefore, we need to see these accommodations the child has made as accomplishments for them in creating a world that makes sense to them.

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Puzzling Behaviors in CHARGE Syndrome

From the FaceBook post:

- Fascinated with fingers/hands (common)
- Laughs at hands
- Plays with hands instead of toys
- Stops what they're doing and touches ground repeatedly
- Knocks on walls/doors before entering a room
- Finger "twiddling" in front of face
- Grinds teeth
- Slaps knees
- Grunts
- Becomes angry when family laughs at jokes/cute things the child says (lunges/hits)
- Violent fits (pulling hair, kicking, punching)
- Constantly touches forehead
- Holds jumper away from stomach
- Leans over to the right
- Bangs elbows on things
- Sticks tongue out a lot
- Touches the inside of eyelids
- Spins in circles
- Bites fingertip with tip of it under tongue
- Obsessively collects things
- Skin picking (lips and fingers)
- Arms crossed
- Lies with one leg over knee
- Chews on things
- Sticks fingers in mouth to the point of gagging
- Spits/puts spit on their hair (or someone else's)
- Spins on floor
- Empties containers/drawers & put everything back
- Hands in fist with thumb tucked in
- Fingers in eyes
- Bangs head on things
- Tongue constantly sticking out
- Fascinated with lights
- Clears bed of anything on it
- Crawls on-top of people lying on the floor
- Flails body when happy
- Hits self in face
- Shakes head from side to side
- Archs back
- Turns on and off lights



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“CHARGE” is Enough



David Brown Tim Hartshorne

23rd June – 4.00 p.m.

***“CHARGE Syndrome and Behavior:
Diagnosis and Intervention (Part II)”***

Interventions for challenging behavior. This session describes interventions that can help to reduce the behaviors that often lead to psychiatric diagnoses. These interventions follow the principle that the behaviors are functional for the child given their various impairments. The interventions address issues of pain, sensory processing, and anxiety.

So what the h_ll are we supposed to do?

Coping with the behavior of people with CHARGE

Tim Hartshorne, David Brown, and Shanti Brown

One way of sorting and categorizing varied behaviors would be to list them in increasing order of urgency for intervention in the form of the following questions:

- 1. Is this a behavior that just bugs you personally, so that it can be accepted and ignored?**
- 2. Is this a behavior that seems to help the child to function in a positive way, so that it can be accepted and ignored?**
- 3. Is this a behavior that seems to help the child to function in a positive way, but should be reduced, or replaced by another, better behavior over time?**
- 4. Is this a behavior that is undesirable and needs to be reduced or replaced fairly quickly?**
- 5. Is this a behavior that needs to be prevented immediately?**

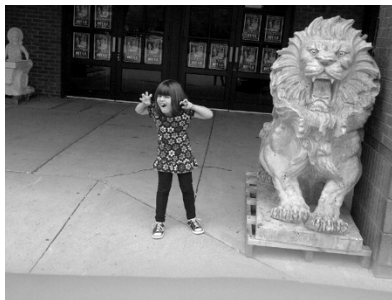
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Jane bangs her head against the side of the bathtub while taking a bath.
Alan flaps his arms and hands wildly when he is excited.
Megan sticks her hands in her food at some meals.
Bill refuses to get out of bed some mornings, but will just lie there and laugh.
April will sometimes press her fingers into her eyes.
John wants to pull the fire alarm whenever he sees one.
Amber interrupts the class with noisy outbursts.
Tim will drop to the ground sometimes when out on walks or excursions.
Hayley will often make clucking noises that go on and on.
Jason often rocks his body while sitting at the table.
Emily pushes people when they are in her way.
Trevor will sometimes hit and scratch people who are near him.
Heather will bite her fingers until they bleed.
Jared repeatedly checks where his cup and plate are at meals.
Nancy likes to hang upside down on her chair or couch.
TJ will put his hands behind him and grab feces and smear it over his face.
Toni likes to throw her toys after she has played with them.
Ray likes to look at lights and will hold a light close to his eyes.
Jacob will play with people's shows and likes to lick the bottoms of them.
Lauren will have a temper tantrum if anything in her schedule changes.



Interventions

- Always consider pain first!
- Figure out why
- Co-regulation – what do you do to stay basically in control?
- Yoga or Tai Chi
- Sensory breaks
- Calendar systems
- Social stories
- Experience books
- Respect communication
- Managing transitions
- Adapt the environment



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PAIN?



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Understand behavior first



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This is normal viewing posture...

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



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Co-Regulation

What works for you in managing

Your thoughts and motivation

Your feelings and emotions

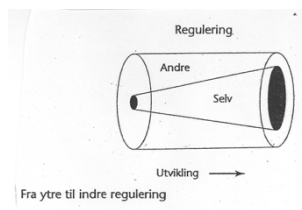
Your actions and behaviors

Your physical state

How do we teach this to our children who do not learn passively?

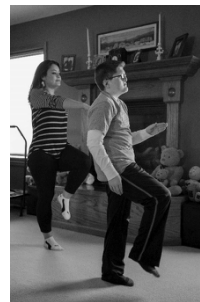
Supporting self-regulation

- Because self-regulation skills are hard for children with significant disabilities to develop
- We provide the external support for what will become an internal self-regulatory process

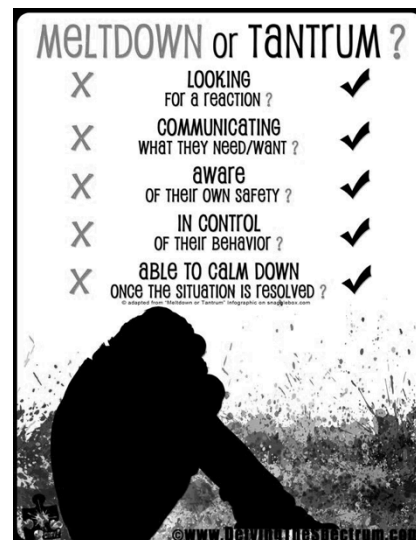


Fun Chi

Reduced stress
Reduced anxiety
Reduced depression
Increased self-esteem
Increased energy/focus/concentration
Increased positive mood
Better balance
Improved sleep
Improved immune system

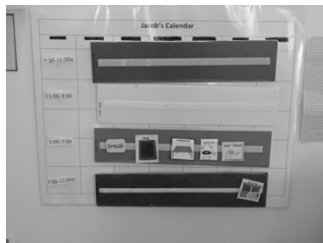


Sensory breaks



Calendar systems

- ❑ Anxiety due to a lack of predictability
- ❑ Utilize an informed calendar intervention
- ❑ Increase self-regulation and predictability
- ❑ Reduce anxiety and anxiety related behaviors



Social Stories

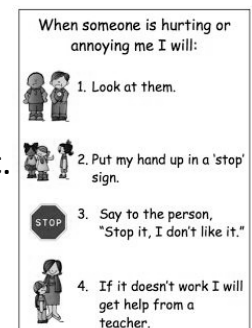
Short, simple, to the point, easy to make, personalized

Conveys social situation and explains how someone would react or what to do

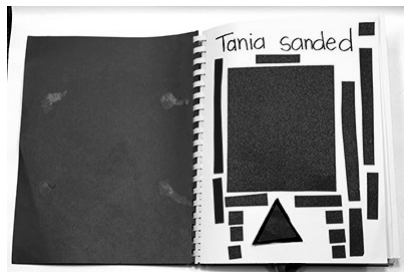
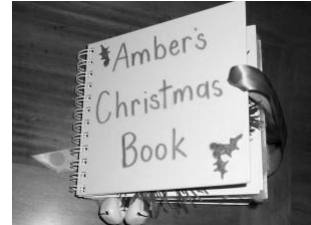
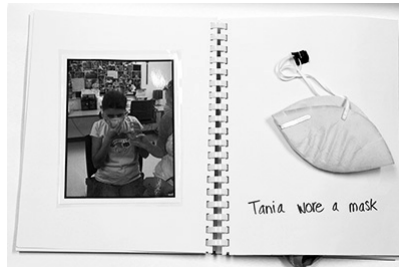
Repetition makes the message stronger

Can help teach a lesson visually

Potty training, sharing, handling anger, change in routine, etc.



experience



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Respect communication

Communication vs. compliance: Jessica

Jessica will only sit in her chair for about 10 minutes at school before she wants to get down.

When she is told that she must stay in her chair longer she tantrums and has a meltdown.

One day she scratched her intervener and then flung herself from the chair and ran to a corner of the room where she lay down on the floor.



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Jacob and the bath



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Managing Transitions

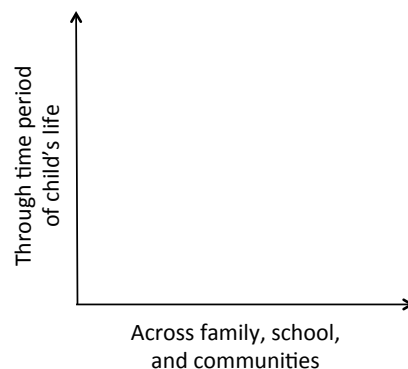
“Goal of a successful transition process is the child’s success in the next environment.” (Rouse, Hallam, Harbin, McCormick, & Jung, 2007)

Types of Transitions

Between daily activities

Life events

Changes in setting



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Managing Transitions

- Visual and auditory cues
- Visual and tactile daily and weekly schedules
- Explicit teaching on procedure for transitions
- Extra time for processing directions
- Reminders
- Social Stories
- Calendar systems
- Visit and explore new settings



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We know that external factors in the environment can have a profound impact on a child's behavior, so one way we can modify what the child is doing is to change features of the environment.



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Key Elements in Environment

- People
- Time
- Space



Tony Best (1998, July) Structuring the Environment. *DbI Review*, 4-9

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People

- Limited in number
- Available
- Responsive
- Not overburdened with non-child tasks
- Consistent
- Engaged
- Skilled observers
- Familiar
- Identifiable (personal markers)

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Time

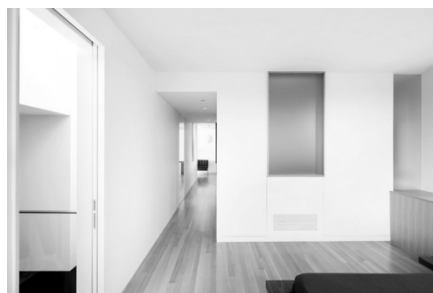
- Sequences
- Consistency
- Number
- Calendars/ schedules
- Survey past, and anticipate & plan future
- Repetitions (with concrete markers)
- Adapted timepieces



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Space/Place

- Markers to identify areas by use/function
- Accessible
- Consistent
- Responsive
- Uncluttered
- Routes
- Landmarks



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Remember that a child's behaviors can indicate what is wrong/missing from their environment, and can also sometimes point you towards the solution to the problem.

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Minimize tactile distractions



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Provide the necessary physical supports for postural security



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Utilize and allow the most effective postures for attention and comfort



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Minimize ALL distractions!



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Keep smell & taste inputs pure and separate



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Position and support for optimum attention, comfort, and functioning



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Managing the environment to prevent behavior problems

Consider what you could do to make the environment of your child safe and secure to enable their optimal development, and to avoid challenging behaviors

- Familiar – how could you create an environment that feels safe and secure to the child?
- Regular – how could you create an environment that is predictable to the child?
- Responsive – how could you create an environment that anticipates and responds to the behavior of the child?
- Meets basic needs – how could you create an environment that anticipates and responds to the needs of the child?

For each question, consider the impact of people, time, and space, and sensory impairment.

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Resources

- Barrey-Grassick. S. (2011). Sign Chi: signing a way to relaxation and stress reduction. *DeafBlind International* 47, 11-13.
- Tony Best (1998, July) Structuring the Environment. *Dbl Review*, 4-9
- Calendars: For Students With Multiple Impairments Including Deafblindness -- Blaha, Robbie. Austin: Texas School for the Blind and Visually Impaired. (2001)
- Lewis, S., & Tolla, J. (2003). Creating and using tactile experience books for young children with visual impairments. *Teaching Exceptional Children*, vol. 35, No. 3, pp. 22-28, Jan/Feb 2003
- Hartshorne, T. S. (2003). Positive behavioral supports and social relationships. *Deafblind International Review*. July-December, Number 32, 4-6.
- Ramirez, M. A., Hartshorne, T. S., & Nicholas, J. (2014, January). Self-regulation in individuals with CHARGE syndrome. *Dbl Review*, 52, 43-47.

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Gail Deuce

24th June – 09.30 a.m.

„Managing behavior in the classroom”


Parents and professionals often talk of behavioural difficulties encountered in individuals with CHARGE. Gail will discuss this with consideration to the CHARGE behavioural phenotype proposed by Hartshorne (2011), looking at potential triggers and how we might respond to this in the classroom.



Managing behaviour in the classroom for learners with CHARGE syndrome

Dr Gail Deuce
Oberwesel, Germany
Germany 2018

1



“Challenging behaviour in CHARGE is not inevitable in individuals with CHARGE syndrome, but it is highly predictable”

(Hartshorne et al, 2017)

2

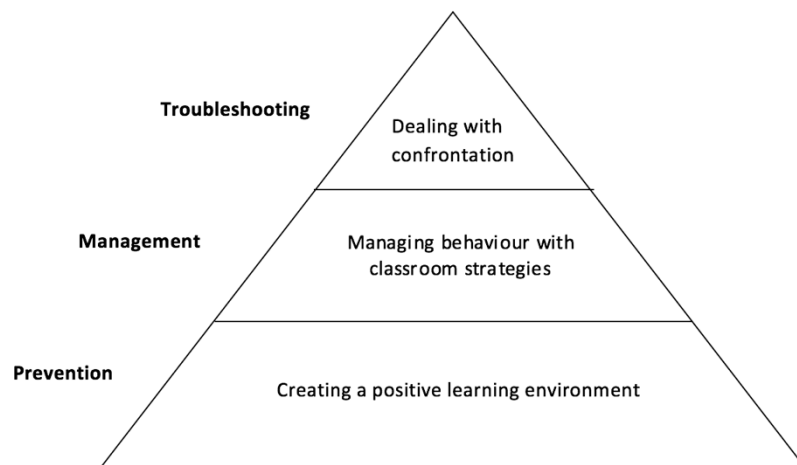
Behaviour in learners with CHARGE

A CHARGE behavioural phenotype (Hartshorne, 2011):

- Low normal cognitive functioning
- Very goal directed and persistent with a sense of humour
- Socially interested but immature
- Repetitive behaviours that increase under stress
- High levels of sensation seeking
- Under conditions of stress and sensory overload, find it difficult to self-regulate and easily lose behavioural control
- Difficulty with shifting attention and transitioning to new activities; easily lost in own thoughts

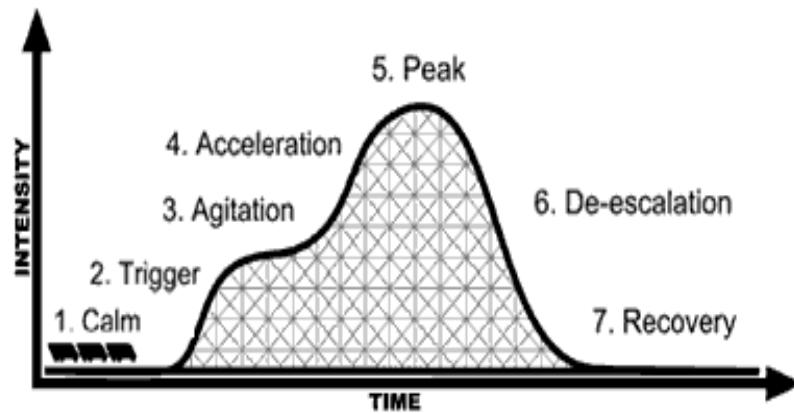
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A model of behaviour management (Birkett, 2016)



4

Managing behaviour in the classroom



5

Possible triggers of difficult behaviour 1

- Pain
- Sensory and physical difficulties
- Increased levels of stress and anxiety, and fatigue
- Difficulties with self-regulation, especially when in a heightened emotional state

6

Possible triggers of difficult behaviour 2

- Previous experiences
- Poor communication skills
- Environmental factors (what is happening around the child/young person)

7

Planning a response

- A need to consider:
- Internal factors
- External factors

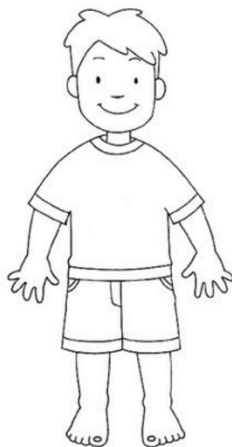
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Managing behaviour in the classroom 1

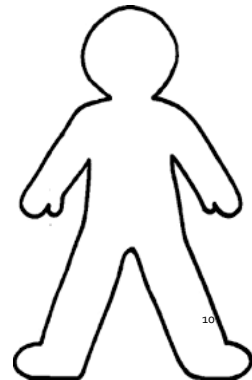
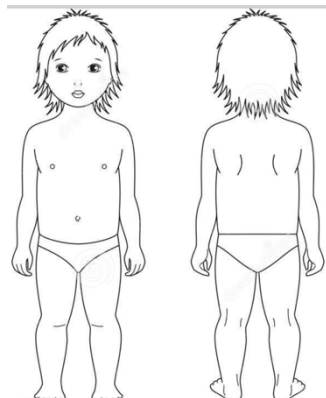
- Teaching strategies to indicate/communicate pain issues and related emotions
- Identify potential sources of anxiety and stress and work to minimise these
- Teach strategies to help manage stress and anxiety
- Accept the learner is likely to need high levels of sensory input/ stimulation. Address sensory issues
- Acknowledge and respect self-stimulatory behaviour and OCD behaviour
- Teach self-regulation strategies
- Identify the child/young person's learning characteristics and how to respond to these (Deuce, 2017)
- Support communication development

9

Communicating about pain- Learning about our body



head
body
leg
neck
foot
finger
hand
arm



10

Communicating pain



11

Managing behaviour in the classroom 2

- Capacity-building model: recognise the learning potential and ability to succeed in school
- Provide predictable daily routine and structure
- Be prepared to share ownership of learning and negotiate
- Know it is not always possible to redirect a child's goals (Hartshorne, 2011)
- Be flexible
- Watch for indicators of emotional and behavioural 'meltdowns' and have a planned response in place
- Give the learner a 'way out'
- Reinforce positive behaviour, including a reward system

12

School rules

Photo



Working at a table



Throwing things



Sharing



Pushing



Shouting

13

Good work: *child's name* 😊



Doing my work



Helping others

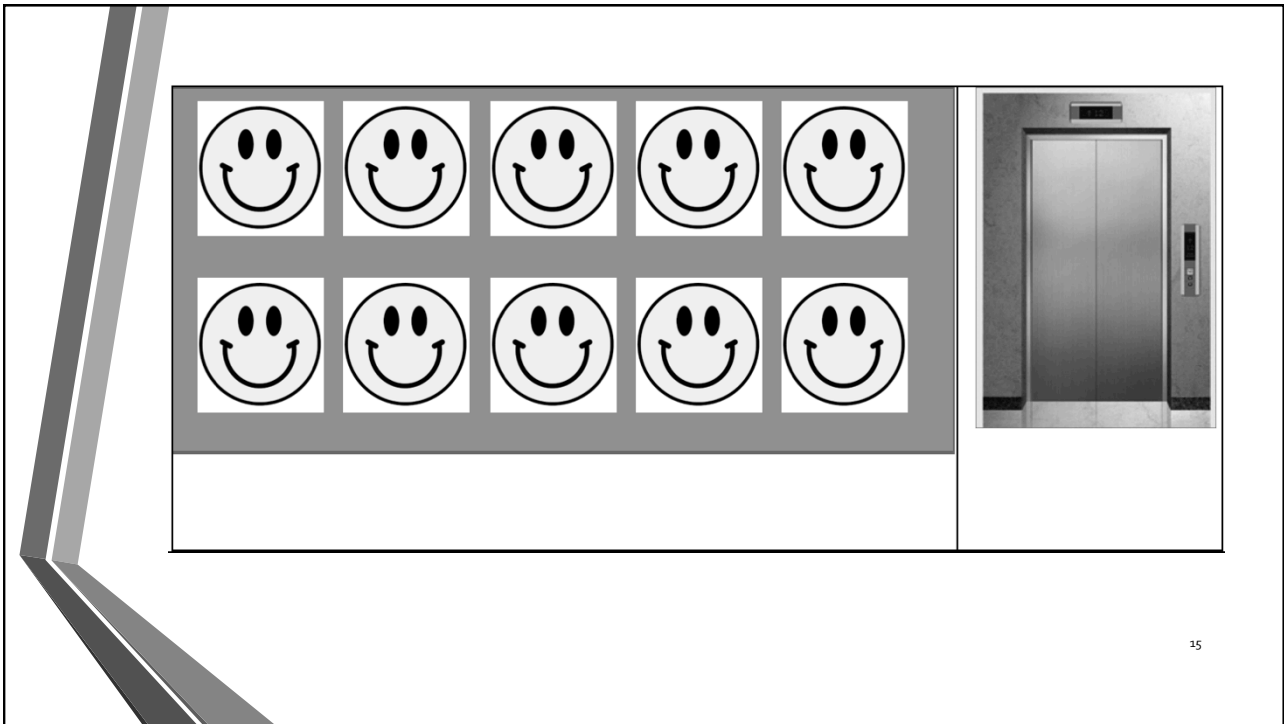


Playing with others
and sharing

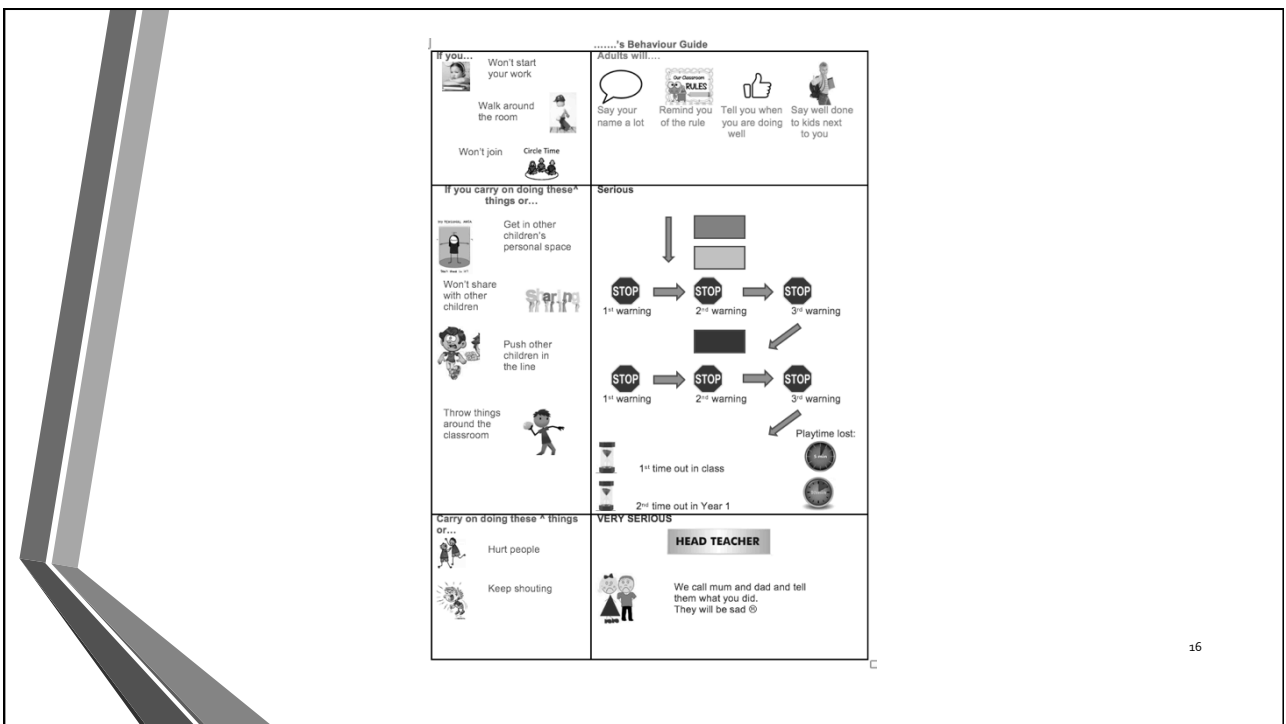


Photo/symbol of reward chosen by child

14



15



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Behaviour support plan

1. Description of *child's name* when she is at each of these stages:

Green Calm and relaxed	Amber Early warning signs, anxious, aroused or distressed	Red Incident!	Blue Calming down Aiming to return to green Reconciliation
<i>Examples</i> E.g. in control, engaged in group activities, playful tone of voice, eye contact, relaxed body language	<i>Examples</i> E.g. agitation, difficulty responding to adult direction, shouting, pulling hair, removing self from group situation	<i>Examples</i> E.g. throwing objects, hitting out at others, shouting	<i>Examples</i> E.g. wanting to say sorry, attempting to re-join group, can accept adult trying to communicate with him/her

2. Strategies for communicating and working with *child's name* at each of these stages:

Green strategies Calm and relaxed <i>E.g.</i> Follow daily routine and structure wherever possible Use visual/concrete supports to prepare him/her for what is to happen next Use first/next board consistently Provide regular movement breaks Allow child to make choices from a selection- sharing control Use reward system to reinforce positive behaviour and increase use for more difficult tasks Catch him/her being good and doing the right thing! Ensure s/he feels a sense of success regularly during the day Feed back to parents about all the good little things	Green behaviour/ preferred activities Calm and relaxed <i>E.g.</i> S/he engages in familiar routines and after anticipating what is to happen next, cooperates Follows adult direction Looks at his/her visual calendar and helps create the calendar for the morning/day. Talks about the activities with an adult Likes to do simple counting worksheets Enjoys ticking off the activities s/he has done Smiles a lot and gives good eye contact Sings
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Amber strategies Early warning signs, anxious, aroused or distressed <i>E.g.</i> Make fewer demands on a difficult day Re-direct him/her using visuals, remind him/her of the class rules Distract him/her with a favourite activity Offer help Lots of praise for the positives Reduce the complexity of the task Ignore the negative behaviours and do not become side-tracked by his/her distractions	Amber behaviour Early warning signs, anxious, aroused or distressed <i>E.g.</i> Sensory seeking behaviours increase Breathing quickens, rubs head and sometimes pulls her hair Shouts and uses the word NO a lot May run away and try to take an adult with her by pulling their hand Changes the subject using objects around her to distract herself and the adults Throws objects
Red strategies Incident! <i>E.g.</i> Ignore the behaviours Keep the child and yourself safe Alert senior management team Following the planned response for a crisis	Red behaviour Incident! <i>E.g.</i> Shouts and screams May kick or hit others May use objects as missiles and cause damage to property Appears extremely distressed
Blue strategies Calming down; Aiming to return to Green; Reconciliation <i>E.g.</i> Allow time to calm down without additional demands Offer a sideways hug When calm refer him/her to their visuals	Blue behaviour Calming down; Aiming to return to Green; Reconciliation <i>E.g.</i> Can be very tearful and apologetic Needs reassurance Can be lethargic and reluctant to do anything Exhausted and difficult to regain their equilibrium

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British Journal of Special Education Vol.44(4), pp.376-393

Hartshorne TS, Stratton KK, Brown D, Madhavan-Brown S and
Schmittl MC (2017) Behavior in CHARGE syndrome. *American
Journal of Medical Genetics* Vol.175C pp.431-438

CHARGE Non-Vocal Pain Assessment (CNVPA)
[https://www.chargesyndrome.org/wp-content/uploads/2016/03/non-
vocal-pain-assessment.pdf](https://www.chargesyndrome.org/wp-content/uploads/2016/03/non-vocal-pain-assessment.pdf)

DBI Review- series of articles on self-regulation in CHARGE

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