CHARGE Syndrom e.V.

Tim Hartshorne

June 25, 2022

„Raising a child who has CHARGE”

Raising a child who has CHARGE

A survival guide

MARATHON SKILLS

Ann P. Turnbull

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

Turnbull, A. P. (1988). The challenge of providing comprehensive support to families. *Education & Training in Mental Retardation, 23*(4), 261-272.

Maslow's Hierarchy of needs

Where are the parents?

Family strengths and needs

- Values
- Coping strategies
- Resources
- Pressure points
- Joys

FAMILY NEEDS

- Information
- Family And/or Social Support
- Accessing Community Services
- Help Explaining To Others
- Financial Assistance
- Child Care

COURAGE IS NOT DENIAL

BUT SOMETIMES COURAGE LOOKS LIKE DENIAL TO OTHER PEOPLE

Professional Misunderstanding and Parent Binds

- Unhappy and discouraged need help
- Happy and optimistic are in denial

- Overinvolved
- Underinvolved

The problem is how to love your child with no need for your child to be any different, without giving up hope that your child may progress with time and resources.

SENSE OF COMMUNITY
McMillan & Chavis (1986)

MEMBERSHIP

INFLUENCE

FULFILLMENT OF NEEDS

SHARED EMOTIONAL CONNECTION

Emotions

As a baby it was fear and worry with all the diagnoses/surgeries and not knowing what was next. Not knowing how well she could see or hear and just observing and waiting. But pure JOY when she hit a milestone!
As a kid stress and worry as she enters school in a gen Ed classroom. Will peers accept her, will she make friends? (they do/she does) STRESS for IEPs and battling with school to get her what she needs to have equal access. This takes so much time and preparation.
Always worry... never knowing what the future holds for her. She is 10 now and I question if she will go to college, or be able to drive, if she will be able to live on her own.

Benefits

From my point of view, now I appreciate every step, simple things. I have different priorities, I try to be more patient, kind. I think there are a lot of benefits. But! I am totally different from a mother who raises only "typical" children.. A little bit weird!!!

Yes! I learned to turn my hurt and anger into Advocacy and both me & my child have benefited from it!

Waiting for the case manager

- I don't know how to do any of this
- I am a bit overwhelmed by all of the booklets, leaflets, handouts, manuals
- All these medical people tell me things and I only understand part, and they don't talk to each other
- I know there are programs and services in my community, but I don't know what they are or how to access them
- There has to be someone who can manage all of this!

Get organized

- Problem solving
- Communication
- Networking
- Collaboration
- Education
- Community organizing
- Persistence
- Trust your gut

Person Centered Planning

Networking

- Identify Needs
- Identify Resources
- Link Resources To Needs

Balancing the life tasks

- Society presents us with three tasks for living
  - Work – we need to be productive
  - Friends – we need to get along
  - Love – we need to commit to family
- We need to find a balance among these

Life Tasks: Parents

- Work – may need to change due to child's needs
  - May also change due to what I have learned

- Friends – may no longer have the friends I once enjoyed
  - May have a whole new set of people who are living my life

- Love – may be a challenge to my marriage and family
  - May lead to a new sense of love and commitment

Good enough parenting

Perfect courage is the courage to be imperfect

And a little bit weird

Contact information

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www.chsbs.cmich.edu/CHARGE

CHARGE Syndrom e.V.

David Brown

June 25, 2022

„What do you do?“
The importance of observational assessment and following the child

"What do you do?": The importance of observational assessment and following the child

16th CHARGE Conference
Oberwesel / Germany
24th – 26th June 2022

David Brown
Deafblind Educational Specialist

The uniqueness (and complexity) of CHARGE syndrome

The changing nature of the population of children with CHARGE syndrome

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."

CHARGE - the most 'multi sensory impaired' of all syndromes
Problems with the perception of:

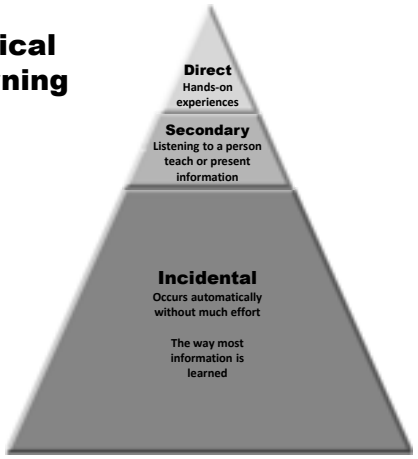
- Vision
- Hearing
- Touch
- Proprioception
- Temperature
- Pain
- Vestibular
- Smell
- Taste

Why do people assess children with CHARGE syndrome?

Challenges to the Assessment Process

- CHARGE presents a very diverse and complex population of learners
- CHARGE presents a wide variety of idiosyncratic behaviors
- People doing assessments usually only know one type of assessment process
- There are limited resources and assessment tools available
- People doing the assessment often forget "The reason why" of assessment

Typical Learning



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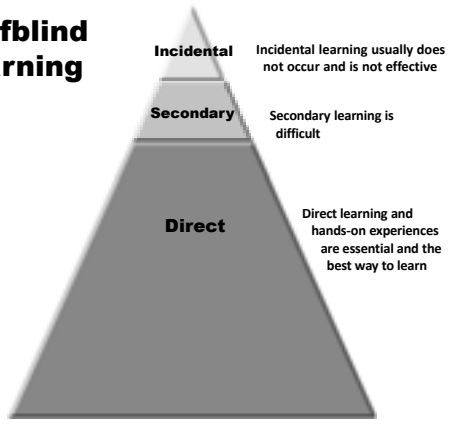
Deafblind Learning



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My view of assessment…..

- Is unusual!
- Is positive
- Looks at positive skills & achievements
- Looks at learning styles
- Looks at preferences & interests
- Looks at the whole child
- Credits the child with intelligence

My view of assessment (2)…..

- Seeks to improve my understanding of the child
- Seeks to help me to build a positive relationship with the child
- Seeks to help me to know what to teach and how best to teach it
- Seeks to give me a clear focus for measuring progress

What might be going wrong with an assessment? (1)

- A focus on deficits rather than skills
- Ignoring the child's motivators
- The pacing is inappropriate for the child, their health, their alertness level, the time of day, the place, the activity, the people present, & the materials used
- The task is too complicated and too challenging
- Too serious - where's the play?
- The expectation that the child will 'Pass or Fail'

What might be going wrong with an assessment? (2)

- Inappropriate assessment tools
- Mimicry of standardized clinical assessment procedures (timing, positioning, materials, expected outcomes)
- Lack of a whole-child perspective
- The assessor is wearing blinkers ("This is what I want to see, now!")
- The assessor is assessing the wrong things
- What is a response?

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

The right educational program for each child with CHARGE syndrome never already exists but must be created. The program must be fitted to the child, not the child to the program.

Jan van Dijk (1966)

In the educational atmosphere I describe, the child holds the central position, the teacher 'follows' the child and, when the child responds, the teacher is present to answer the child's request

The Van Dijk Approach - Evaluation challenges

- No prescribed protocol
- No specific implementation order
- No set of testing materials
- Each assessment is unique
- No set interpretation scale

The Van Dijk Approach to Assessment

- Child-guided
- Fluid
- Looks at the processes children with multiple disabilities, including sensory impairments, use to learn & to develop
- Assessment is summarized in terms of strengths and next steps for intervention

Basing the assessment approach on the child's curiosity and personal satisfaction, on current abilities and interests rather than on current deficits, on function rather than on structure, on motivated behavior rather than on sterile performance, is now seen as a legitimate and effective way of beginning the process. The approach needs to be individualized and holistic, so that every aspect of the child is taken into consideration even if only one sensory or skill area is being assessed. The emotional needs of the children will exert a direct and powerful influence on their ability to function, so that serious consideration of questions like "How do you feel?" "What do you like?" and "What do you want?" will provide the best basis for successful assessment. People often think that "What can you do?" is the key question to pose to any child during an assessment, but with this group a better question to begin with would be "What do you do?"
D Brown "Follow the child" reSources Vol 10 No 9 Winter 2001

Assessment Questions
D Brown "Follow the Child" (1999)

- How do you feel?
- What do you like?
- What do you want?
- What do you do?

How do you feel?

*

Arousal/ bio-behavioral state (Availability for learning)

"Availability for learning refers to the time when an individual is paying attention, and is able to listen and respond…. Many children with sensory impairments…. have medical and health challenges. These factors often make self-regulation particularly difficult. A Biobehavioral State is a state of arousal."

Chris Russell
https://www.pathstoliteracy.org/resources/biobehavioral-states-and-availability-learning

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-stimulation & Self-regulation

What do you like?

*

Motivation & Meaning

What do you want?

*

Current expressive behaviors

What do you do?

*

Spontaneous, without and before intervention

Most people focus on the child's disabilities, but close attention to their abilities can reveal more about the difficulties they face as well as the strategies they use to function effectively.

Everything that children with CHARGE syndrome do has meaning, and the first obligation on the teacher is to ascertain that meaning (or at least to come up with a really good guess).

CHARGE Syndrom e.V.

Kim Blake

June 25, 2022

„Missed medical diagnosis in CHARGE syndrome – POTS, migraine, cyclical vomiting and more”

Missed medical diagnosis in CHARGE syndrome

POTS, Migraine Cyclical vomiting and more

CHARGE Syndrom e.V.

Oberwesel
Germany 2022

Dr. Kim Blake, Professor of Pediatrics
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DALHOUSIE UNIVERSITY

IWK Health Centre

Flying again to conferences

Halifax from the air

Trick or Treat from the IWK hospital

Objectives

- To make you aware of Postural Orthostatic Tachycardia Syndrome (POTS) and its treatments.
- To help you understand migraine, cyclical vomiting and their connection to CHARGE syndrome.
- To explain and expand the use of the CHARGE syndrome checklist.

Have the checklist available for later in the presentation

Case report

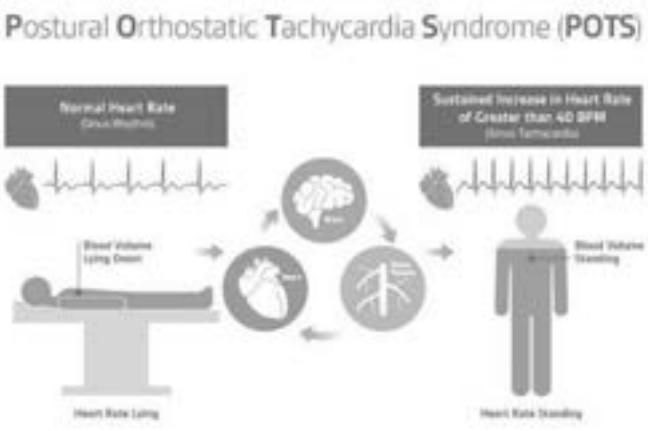
A 17-year-old female with CHARGE Syndrome was seen in the CHARGE Clinic at the IWK with symptoms suggestive of POTS. These included:

- Physical fatigue on hot days over the last year
- Heart rate that at times elevated to 140 bpm
- Agitation and discomfort when moving from a lying to standing position
- Many events of vomiting, loss of colour in the face, sweatiness, pre-syncope and diarrhea following standing up.
- One instance severe enough that prompted a visit to the Emergency Department and admission overnight.

What is POTS?

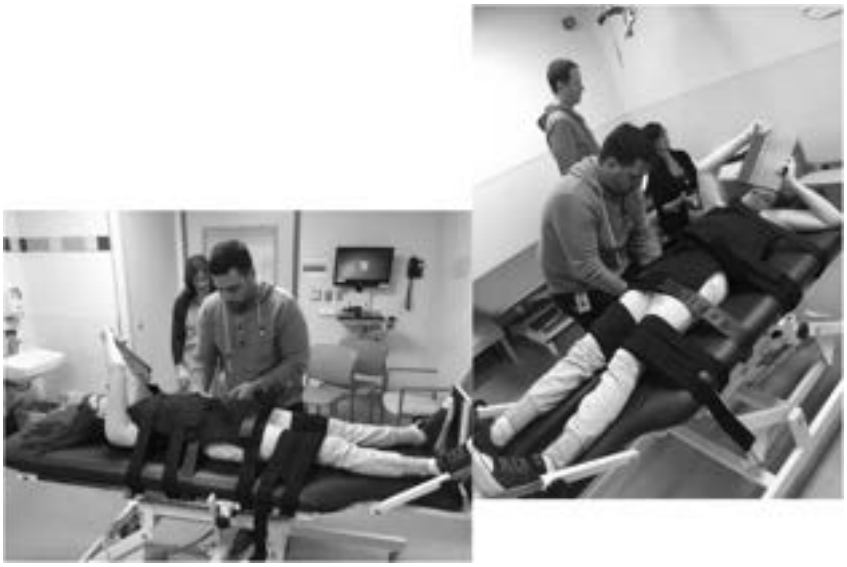
- "POTS" stands for Postural Orthostatic Tachycardia Syndrome
  - Postural = position of your body
  - Orthostatic = standing upright
  - Tachycardia = very fast heart rate
- It is a clinical syndrome that falls under the umbrella of dysautonomic conditions
  - This means that the symptoms arise from dysregulation of the autonomic nervous system

Why does POTS happen? (Physiology)



- POTS is characterized by a rapid increase in heart rate upon standing. Symptoms are often relieved by lying down.
- Vasodilatation in the lower limbs (blue color) not enough blood goes to head and neck. Resulting in increased heart rate.

How to diagnose POTS with a Tilt Table Test



Heart rate and blood pressure were recorded throughout the test. The blood pressure did not change, but the heart rate increased by over 40 beats per minute (76 ➡ goes to 120) when she was positioned to 90° (upright) Lowering her to a supine position returned the heart rate to normal.

Study: Postural Orthostatic Tachycardia Syndrome (POTS) in CHARGE syndrome

Julia Morrison, George Williams, Angela Arra, Kim Blake

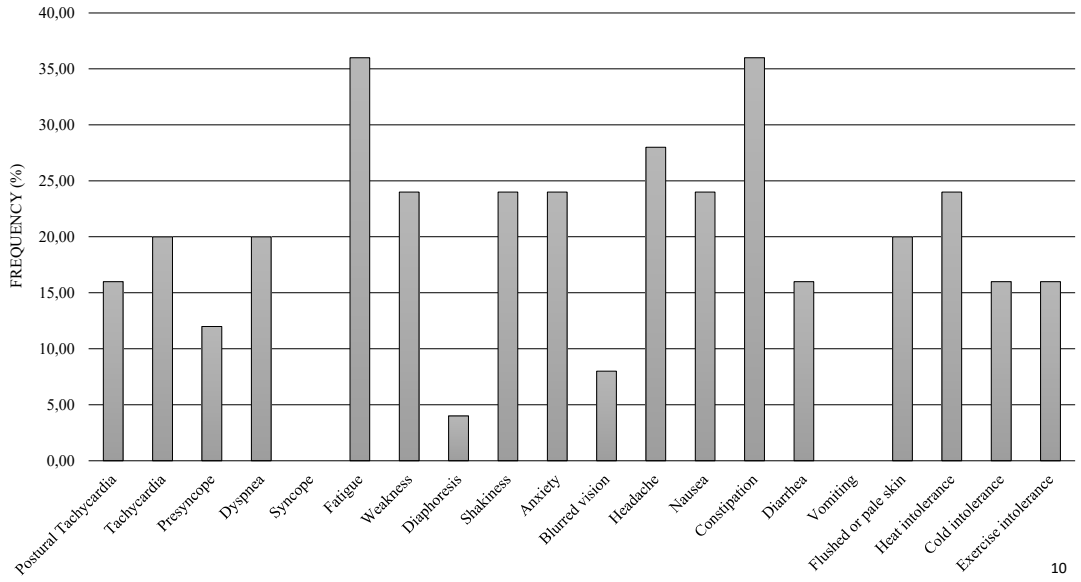
METHODS: The COMPASS 31, a validated questionnaire designed to measure dysautonomic symptoms, was adapted for this study. Data from individuals with CS age 12 or older (not just those with dysautonomic symptoms). The questionnaire listed symptoms (e.g., tachycardia, presyncope, fatigue, nausea, constipation) with a Likert scale from 1 (occurs never) to 4 (occurs often, e.g., once per week)

IWK Health Centre

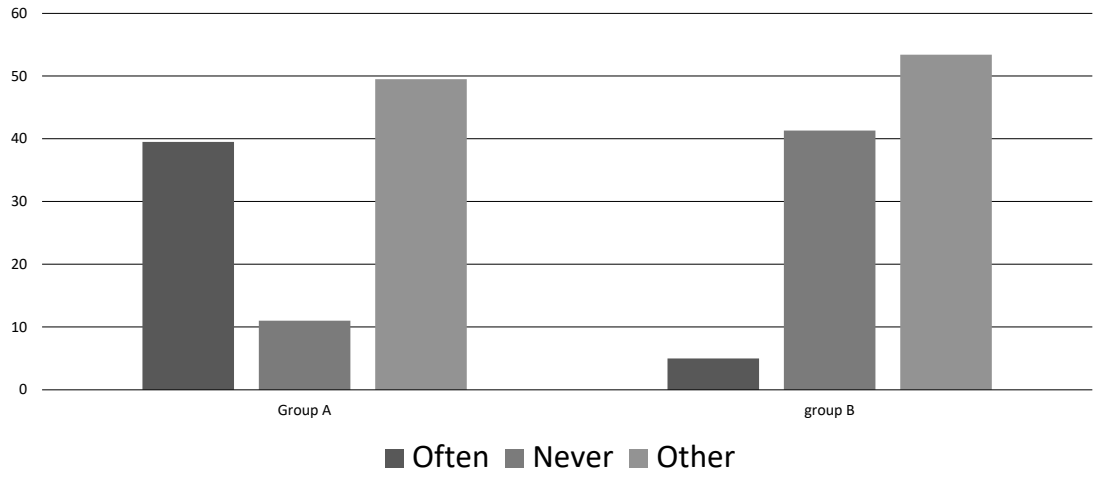
Results

- 25 (F=20, M=5) participants, age range of 12 to 33 (mean 21.5) years.
- Tachycardia, headache, fatigue and constipation (frequency 20-36%).
- The mean number of symptoms occurring **"often"** (ranked 4) was 3.75.
- Responses were stratified into two groups based on this mean. In the "above average" group, (n=10) POTS symptoms occurred with a high frequency (e.g., at least once per week) in 39.5% of participants. In the "below average" group (n=15), symptoms were reported with a high frequency just 5% of the time.

Frequency of Symptoms on a weekly basis



Average percentage of participants in each group who experienced symptoms “often” (weekly) vs. “never”



Group A = above average POTS Symptoms (n=10)

Group B = below average POTS Symptoms (n=15)

In their own words

“…At this point we were using a wheel-chair every time we went to the store because she was so fatigued just from walking to the car to the inside of the store and was getting dizzy.”

- “It’s awful & we have to go to the hospital regularly for fluids to help her come out of the episodes. She can't walk, talk, eat, or drink & feels like she's falling even when she is laying down. She misses 2-3 days of school a week & sleeps most of the time she is there.”

Discussion from our research

- POTS may be more frequently found in older individuals with CS and should alert families and practitioners to consider this when an individual with CS presents with fatigue and dizziness especially when triggered by heat and stress.
- 10-minute stand test can be used in an office setting or at home.
- Dysautonomia; problems with the autonomic nervous system, (ANS) is likely in CHARGE syndrome given the temperature and pain variability.

Postural Orthostatic Tachycardia Syndrome (POTS)

Just like CHARGE syndrome POTS can affect most systems of the body

Symptoms and signs:

- Brain fog (lightheaded ness)
- Fainting
- Heart palpitations
- Fatigue

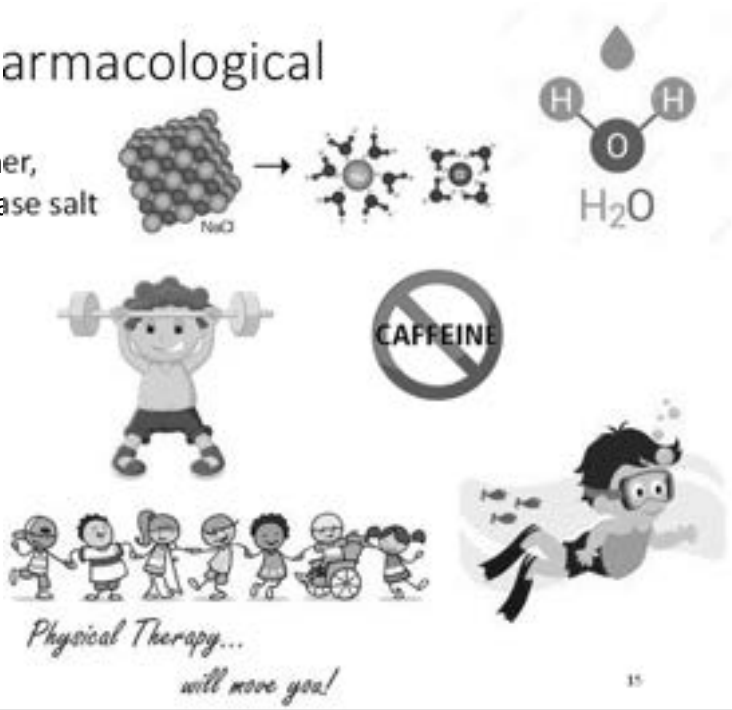
CHARGE conference 2019

Abdominal Symptoms:

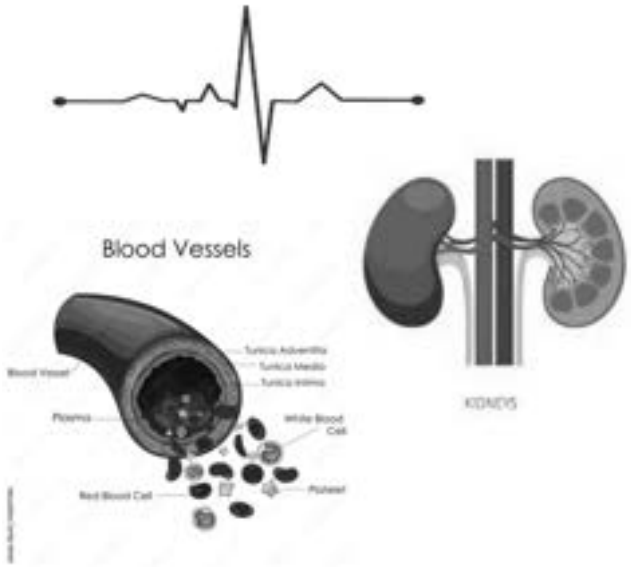
- Vomiting
- Nausea
- Diarrhea
- Bloating
- Constipation
- Cramping

Treatments – Nonpharmacological

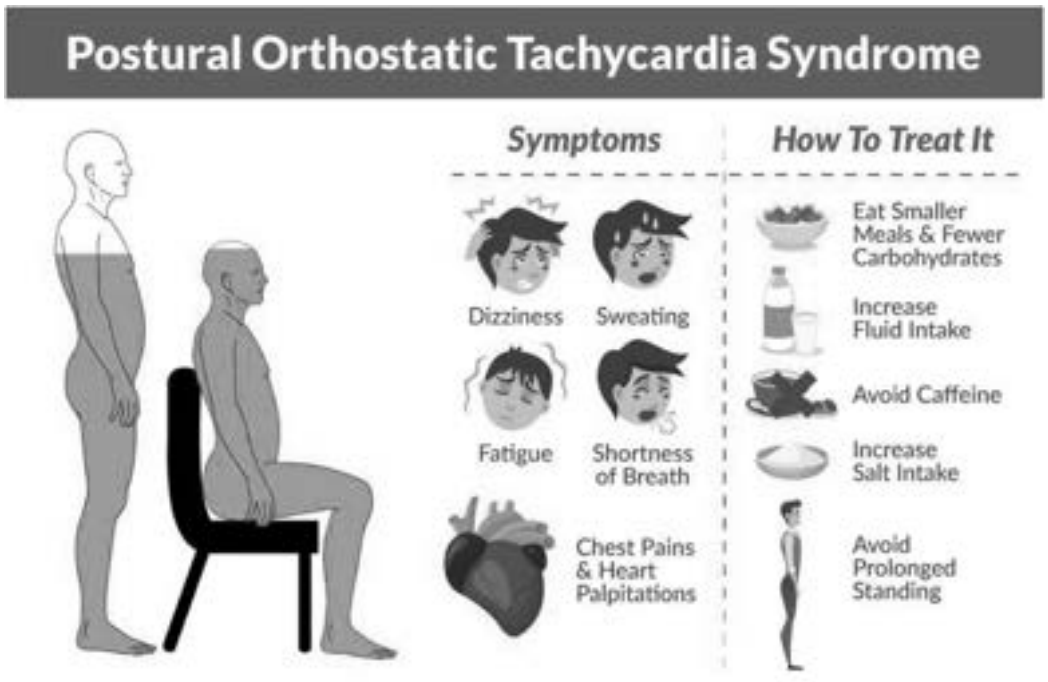
- Watch for triggers e.g., hot weather,
- Fluids with electrolytes and Increase salt in diet
- Avoid prolonged standing
- Limit caffeinated drinks
- Small frequent meals and lower carbohydrates.
- Compression stockings
- Refer to rehabilitation specialists
- Aquatic Therapy for PT – range of motion
- Physical therapy – "build up" conditioning of exercise



Treatments - Pharmacological



- Fludrocortisone. Helps the kidneys retain sodium (to treat Syncope and hypovolemia)
- Beta-Blockers Helps to block effort of adrenal hormone on lowest heart rate. (use low dose for palpitation).
- Pyridostigmine particularly for constipation.
- Midodrine – improving blood vessel constriction.
- Refer to neurology and or Cardiology.



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Summary and Recommendations

POTS is difficult to diagnose – especially with individuals who have multi-sensory impairment

There is likely more overlap with CHARGE syndrome than we currently understand

Awareness of the condition and associated triggers. Monitoring fluid and salt intake, exercise and conditioning and taking breaks during periods of activity

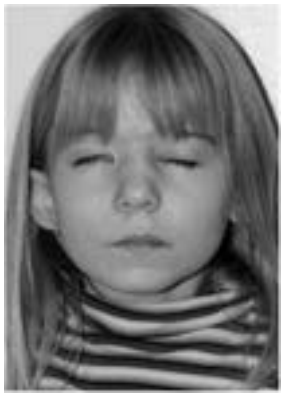
18

Migraines in CHARGE Syndrome

- Can be an atypical presentation in CHARGE syndrome
- Look for family history and triggers.
- "Abdominal migraine" in younger individual can progress to typical migraine

An individual with CHARGE syndrome – presenting with decreased sensitivity of the cornea

- Admitted with diarrhea and vomiting.
- Abnormal ears and facial palsy.
- "Sand not felt in her eyes".
- CHD7 anomalies, diagnosed with CHARGE Syndrome with few clinical features.
- Started to get migraines at age 4 years
- Migraines so severe she would miss school, need to wear sunglasses at all times, nausea and vomiting.



Showing Right facial palsy and Hypoalgesia (Cranial Nerve VII and V)

A case report of Migraine Treatment in CHARGE syndrome using Onabotulinum Toxin A .

Morrison, Fisher, Arra, Bezuhly, Blake

- MT was getting bullied regarding her facial palsy and received injections of Botox® into the left lower lip muscle (normal side) to relax the face for more symmetry
- Coincidentally, this relieved her previously treatment-resistant chronic migraine headaches
- lower lip muscle injections are not typical for Botox in migraine management

www.drkimblake.com

Method of Botox treatment

- Botox treatment to normal facial nerve. Every 3-4 months. Tracking improvement by diary of migraines.
- Possibly practice-changing approach to migraine treatment for children, specially those with CHARGE and/or facial nerve palsies

Migraines – Evaluation and treatment

- Communication challenges
- Atypical presentation of the migraines
- Suggest using Pain Scale and looking for triggers.
- Medical prophylactics treatment – daily vitamin D 1000-2000 units, Magnesium and riboflavin.

Cyclical Vomiting

Four phases

- Normal
- Prodrome – odd feelings
- Vomiting
- Recovery
- Often a history of migraines.

Case history of cyclical vomiting

"CV is rare, CHARGE is rare – don't chalk everything up to its just CHARGE"

- 4-year-old girl with vomiting every 1-2 months for 3-6 days.
- Morning stomach cramping not revealed by anything – venting g-tube, meds – lead to vomiting.
- Family history of sever migraines.
- Investigations normal and no treatment helped.
- Triggers were sleep problems, stress and some additives in junk food.
- Similar management to migraines. Try low fat, more meals with L-Carnatine and Co-enzymes Q10.
- Drugs that can be used are associated with Amitriptyline (or other anti depressants) Valproate, Topiramate, Gabapentin and

CHARGE Clinical checklist:

Health supervision across the lifespan

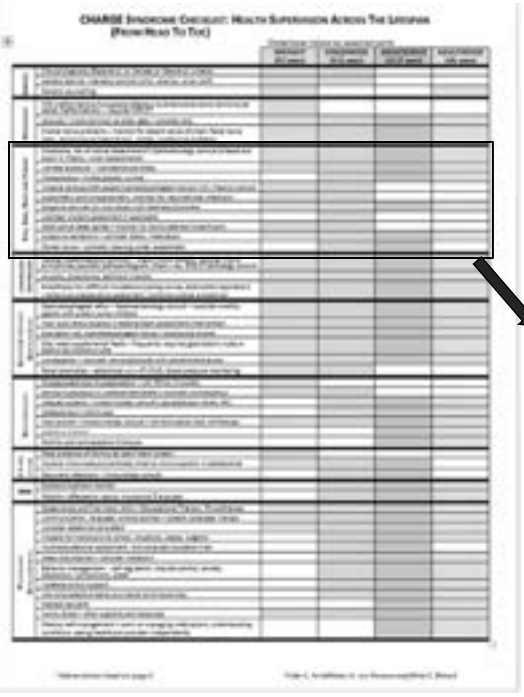
- Key assessment/reassessment points across the lifespan
- Supports all care providers to manage patients with CHARGE syndrome
- Families can use this and bring it to their doctors' appointments.

Trider et al AJMG 2017Mar;173(3): 684-691

Why do we need a CHARGE **clinical** checklist?

- CHARGE syndrome is a complex multi factorial condition and a checklist can…
  - Improve care for individuals and families
  - Prevent missed diagnoses and facilitate early referral
  - Educate learners and professionals who are not familiar with CHARGE syndrome

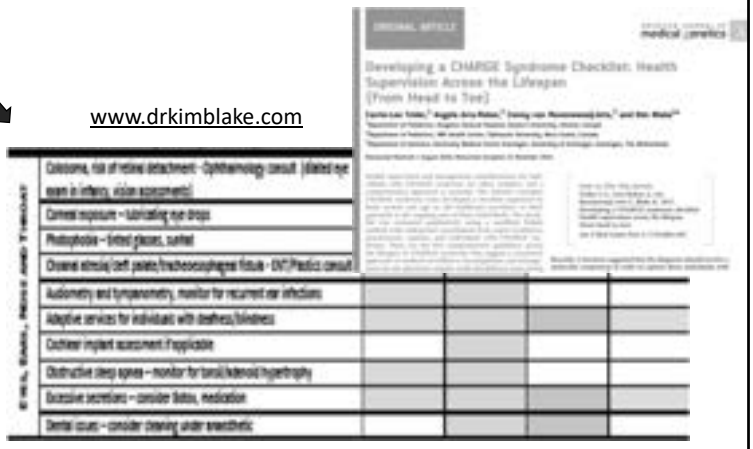
2017 Orlando CHARGE conference



Developing a CHARGE Syndrome Checklist: Health Supervision across the lifespan

(From head to toe)

www.drkimblake.com



Trider et al AJMG 2017Mar;173(3): 684-691 . DOI:10.1002/ajmg.a.38085

How was the CHARGE syndrome clinical checklist developed

- Literature review
- Expert opinion
- Evaluated qualitatively (n=97) by a Delphi method – with Pediatricians, parents, individuals with CHARGE syndrome, deaf blind specialists, therapists =OT PT SLP endocrinologists
- Piloted by 7 multidisciplinary CHARGE syndrome clinics and medical students who had no prior knowledge of CHARGE syndrome.

Trider et al AJMG 2017

Evaluation of the checklist

Guidelines in CHARGE Syndrome and the Missing Link: Cranial Imaging

"CHARGE Syndrome clinical checklist is a well supported frame work for clinical surveillance as demonstrated by a thorough literature search"

- This is the only clinical checklist that has been validated for use with CHARGE syndrome.
- This Guidelines paper includes recommendations for cranial imagining that provide optimal care while limiting risky anesthetic procedures.

RESEARCH REVIEW

WILEY

Guidelines in CHARGE syndrome and the missing link: Cranial imaging

Christa M. de Geus | Rolien H. Free | Berit M. Verbist | Deborah A. Sival | Kim D. Blake | Linda C. Meiners | Conny M. A. van Ravenswaaij-Arts

American Journal of Medical Genetics 2017

Summary of why you and your health care providers should use the CHARGE clinical checklist

- Important issues that professionals need to address at each visit-Age dependent.
- Rare health issues that can be missed in CHARGE syndrome and can lead to adverse health consequences.
- The publication can be used to support the checklist

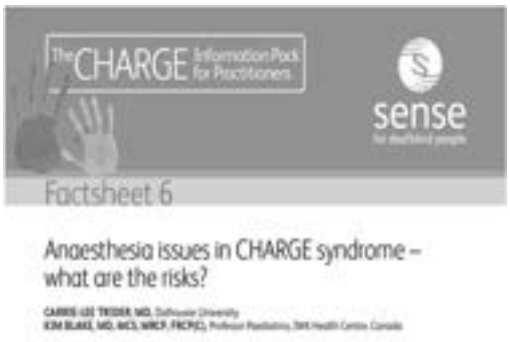
"The CHARGE Syndrome checklist is an important tool for medical students and doctors. It helps them know what questions to ask me and my mom."

" It is nice when I go to a hospital or appointment, and someone has heard of CHARGE Syndrome before!"

Infancy (0-2 years)

Procedures requiring anesthesia should be combined where possible as there is a greater risk of anesthesia complications in individuals with CHARGE syndrome.

https://www.sense.org.uk/get-support/information-and-advice/conditions/charge-syndrome/



Childhood (3-11 years)

Gastrointestinal and feeding issues are prevalent in CS.

- We suspect that lower cranial nerve anomalies (IX, X, XI) produce abnormal gut motility and issues with the gut microbiome.
- Issues of swallowing, reflux, aspiration, abdominal pain (use the pain scale) constipation.
- Multi disciplinary team including pediatrician, gastroenterology, ENT, nutritionist, OT/PT/SL, psychologist.

ENT issues

- Obstructive sleep apnea and consider tonsillectomy and adnoidnactomy

Adolescent (12-17)

Jenna with Micaela 2021

Puberty

- Often delayed / disordered and requires, hormone replacement therapy.
- Referral to endocrinology around age 10.

Bone health is often forgotten

- High dose of vitamin D weight bearing exercises and HRT.

Mental Health

- *Especially Anxiety and ADHD*

2017 Medical student receiving feedback with Jenna

18 years to adulthood

If new behavioral changes arise always consider a thorough history and physical looking for underlying medical issues.

- Hearing and vision i.e. retinal detachment, wax in ears.
- Heart arrhythmias.
- Bloating and constipation.
- Migraine
- POTS (Postural orthostatic tachycardia syndrome).

Obesity is prevalent and its important to involve a nutritionist and physical/recreational therapist.

Where to locate the CHARGE clinical checklist

There is a German translated CHARGE Checklist

www.drkimblake.com

Knowledge translation

International Collaboration is a key to success in a rare condition.

- Website www.drkimblake.com -- a repository of Dr. Blake's research
- CHARGE Syndrome textbook second edition.
- The Atlantic Canadian CHARGE organization. Provides international consultations, problem solving, and support as individuals and families grow up with CHARGE Syndrome.

CHARGE SYNDROME AUSTRALASIA

Online course: Understanding CHARGE Syndrome

Understanding CHARGE syndrome is a 6–8-week **free** online course via an innovative training tool called MOOC (massive open online course). The course is open to anyone in the world with an interest in CHARGE syndrome:

- People living with CHARGE syndrome, their families and careers.
- Medical and nursing professionals
- Allied health professionals
- Teachers and educators
- Advocates, service delivery staff, support workers, planners

https://www.chargesyndrome.org.au/onlinecourse

Thoughts and Questions

CHARGE

CHARGE Syndrom e.V.

Gail Deuce

June 26, 2022

„Supporting the child, young person or adult with CHARGE to make choices and decisions”

Supporting the child, young person or adult with CHARGE to make choices and decisions

German CHARGE conference

June 2022

Dr. Gail Deuce

Preferences

Choices

Decisions

Why is it important?

Research shows, increased choice-making/ decision-making supports:

- Higher quality of life outcomes
- Increased cooperation and task engagement
- Self-regulation of behaviour
- The development of communication skills and social interactions
- Increases independence
- Self-determination
- A person to feel empowered
- A sense of autonomy

Through supporting the development of choice-making/decision-making we can enable the child, young person or adult to make informed decisions and have a voice that is listened to and acted upon

Supporting the development of choice-making/decision-making 1

- Identify preferences (objects, people, places, etc.) and respond to these
- Increase preferences by pairing something preferred with something less preferred
- Build in choice-making opportunities throughout the day
- Make the choices ‘real’, so the child experiences the consequences of those choices/decisions
- Where appropriate, give time and opportunity to change their minds if their choice/decision is ‘unwise’
- Provide rules ahead of time for when choice might not be available or appropriate
- Provide appropriate alternatives

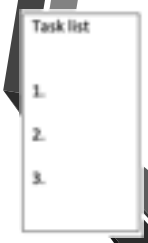
Supporting the development of choice-making/decision-making 2

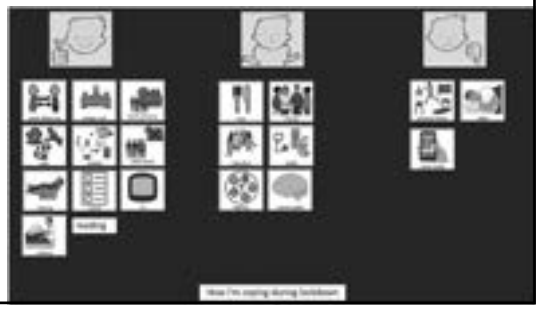
- As choices/decisions become more complex, give as much time and information as possible to help them arrive at their own decision
- Draw on real-life experiences and the experiences of others to broaden their understanding of options, etc
- Respect their choices as different from your own and not just right or wrong
- Where they really lack capacity, give some choices to make that ensure the decision made for them will be in their best interests
- If they (or you) find it difficult to express a choice/decision that is different to your, seek someone else to mediate/advocate

Providing choices at home and in the classroom:

- Activities or tasks to complete
- The order in which tasks are completed
- What materials to use for an activity
- Where to undertake a task or activity
- With whom to undertake the task or activity (friends, supporting adults)

How to communicate their choices/decisions





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CHARGE Syndrom e.V.

Julie Maier

June 26, 2022

„Building a solid educational program: Connecting child-centered Assessment to meaningful learning goals”

Building a solid educational program: Connecting child-centered assessment to meaningful learning goals

Julie Maier, MA
California Deafblind Services
16th CHARGE Syndrome Conference
Oberwesel, Germany June 26, 2022

Agenda

- Linking assessment results to program planning
- Authentic, child-centered assessment
- Developing learning goals
- Considerations for program planning for children and youth with CHARGE syndrome
- Questions

Complexity of educational needs for children with CHARGE syndrome

- Rare condition and most educators have no knowledge or experience with a child with CHARGE.
- Health and medical concerns affect schooling.
- Delayed developmental milestones in communication, motor, and social skills.
- Multiple sensory systems are impaired.
- An educational program must be multi-faceted and use a collaborative team approach.

Jan van Dijk (2001–

"The multi-sensory impaired person is a unique human being with a unique line of development, who is more dependent on the professional's willingness to accept this and act accordingly than any other group of disabled persons."

Linking assessment to program planning

Assessment Process — Complete child-centered assessment to identify strengths, skills, and unique support needs and discuss results.

Learning Goals — Team develops reasonable and relevant goals to build upon current skills and develop new high-priority skills.

Program, supports, and services — An educational setting and necessary accommodations, supports and services are selected based upon the learning goals.

Purpose of assessment

"We assess because there are things we don't know but we want to find out."
-David Brown (2014)

A capacity-building perspective

To develop meaningful educational goals and a program for a child with CHARGE syndrome, you need to identify:

- Child's strengths and skills
- Child's interests and motivators
- How EVERY sense functions and/or is compromised
- Child's communication mode(s)
- Purpose of "behaviors"
- Effectiveness of current supports and accommodations

Key Points of Authentic Assessment

- Assessment is the starting point.
- Family involvement in the assessment process is essential.
- Gather information through interviews with people who know the child well, informal and structured observations, and evaluations by specialists.
- Specialists must collaborate to see "the whole child".
- Assessment of children with CHARGE syndrome must go far beyond the use of typical educational assessment.
- **Standardized tests** may be necessary to qualify a child for services but **are inappropriate as tools to guide educational planning.**

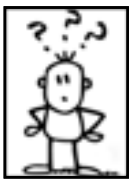
What is child-guided assessment?

Child-guided assessment for children with multiple disabilities, also known as the van Dijk Framework, follows the lead of the child as the assessor(s) attempts to discover the processes a child uses to learn rather than what the child knows. The child is assessed within natural and familiar routines. This approach identifies and builds upon the child's strengths and interventions are developed based upon the findings of the assessment.

(Nelson, van Dijk, Mc Donnell, & Thompson, 2002)

The Four Magic Questions (D. Brown, 2001 & 2014)

- How do you feel?
- What do you like?
- What do you want?
- What do you do?



Critical areas of assessment for individuals with CHARGE syndrome

- Medical
- Sensory
- Communication
- Developmental
- Behavioral

Must include consideration of social relationships and social skill development in these 3 areas.

Source: (Slavin, L.A., 2018. *A Checklist of Educational Needs for Children with CHARGE Syndrome*.)

A Checklist of Educational Needs

The checklist is a tool that educational professionals can use to identify the child's current needs and plan programming.

The checklist links a CHARGE characteristic or concern to:

Educational Needs	Team members to consult	Examples of Strategies and Accommodations	Consulting professionals to provide training and support

Source: (Slavin, L.A., 2018. *A Checklist of Educational Needs for Children with CHARGE Syndrome*.)

Assessment Detours

- Identifying developmental disability without considering vision and hearing loss.
- Identifying autism or attention deficit disorder without considering features of CHARGE syndrome that are similar.

- Neglecting to consider the influence of the environment on the child's performance (lighting, position, physical supports, sounds, familiar materials).

Important areas for assessment

- All 7 senses
- Biobehavioral state regulation
- Communication (includes "behaviors")
- Concept development
- Self-care and daily living skills
- Orientation & mobility
- Personal-identity & self-image
- Academics and use of assistive technology
- Social/emotional needs

Who should be involved in assessment?

- Family.
- Familiar and trusted teachers and support staff.
- Specialists with expertise in areas being assessed (vision, hearing, language, behavior etc.).
- Ideally, someone with knowledge and experience of deafblindness or CHARGE syndrome.
- May need consult from medical professionals.

Planning & Gathering Information (before assessment begins)

- What do you want/need to know?
- What information do you already have?
- What information do you still want or need?
- Who else might have this information?
- Gather important medical and developmental information prior to assessment from family and current teacher(s).

Holistic sensory approach

- A good assessment will consider presence and use of all seven senses, including proprioceptive, vestibular, smell and taste.
- Attend to:
  - How the senses efficiently work together.
  - How one sense might interfere with another
  - How one sense may compensate for another sense
  - What information is being sought, gained, or avoided

Behavior = Communication

Unusual and unexpected behaviors are often misunderstood and labeled "attention-seeking". Perhaps a better description is **"connection-seeking".**

- **External communication intent-**
  - Connecting and responding to others
- **Internal communication intent-**
  - Connecting and responding with your body or senses

Important to recognize the difference and respond accordingly.

What to look for in assessment results

- Sensory functioning and sensory needs.
- Child's motivators.
- Emerging skills in communication, motor skills, academic skills, social skills, life skills.
- Current supports:
  - What's working?
  - What's not working?
  - What's missing?
- Purpose of behaviors:
  ✱ Attention ✱ Access ✱ Escape ✱Sensory

A complete, child-centered assessment will identify:

- The child's strengths
- The child's interests or motivators
- The child's current and emerging skills
- Necessary supports and services
- Effective accommodations and supports
- Potential learning goals to target

Example: Jessy, a preschool student

- Loves music, dance and taking photos with iPad
- Total communication approach (sign language, speech, photos)
- Supported seating increases focus
- Needs frequent breaks to lie down and rest
- Transition items help her follow class schedule
- Hates to make mistakes. Show her samples of completed projects.
- Intervener support for health and communication needs and pre-academics support.

Example: Madeline, 1st grade

- Imaginative. Loves to build with blocks and Legos and roughhouse with brothers and peers.
- Needs frequent movement breaks to stay regulated.
- Uses iPad camera to enlarge visual materials.
- Needs more time to complete assignments. Modify amount of work, slow the pace of instruction.
- Hates sudden endings and changes! Alert her before transitions and schedule changes.
- Mimics others. Include peers as partners.
- Intervener needed to explain and modify work, manage behaviors, provide language access.

Example: Timothy, High School

- VERY curious about people and things in every environment he enters.
- Loves to learn and needs modified curriculum (amount and extended time).
- Likes to follow expected routines and know what's happening next.
- Identify certain times to converse about topics of interest.
- Family and school team are focused on expanding his social circle and encouraging independence
- Creative about finding different positions and postures for focusing.
- Nurse, fluent in ASL, serves as intervener for language access, academic and social support, and potential health issues.

Linking assessment to program planning

Assessment Process	Complete child-centered assessment to identify strengths, skills, and unique support needs and discuss results.
Learning Goals	Team develops reasonable and relevant goals to build upon current skills and develop new high-priority skills.
Program, supports, and services	An educational setting and necessary accommodations, supports and services are selected based upon the learning goals.

Developing learning goals

- Address each area of need identified in assessment results.
- Must be achievable, observable, and measurable.
- Include context for when and how it will be taught and practiced.
- Include accommodations and considerations of the child's multi-sensory needs.
- Identified as an important goals by the family and the child, if they are able to let you know.

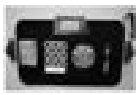
Program considerations for students with CHARGE syndrome

Elements of a strong program

- Accessible classroom and school environment
- Language and communication access
- Meaningful curriculum
- Sensory needs
- Social relationships
- Proper support

Behavioral needs are addressed by careful planning and consistent implementation in each area.

Accessible classroom and school environment



- Organized and predictable.
- Schedule or calendar system is used.
- Includes spaces for sensory break or movement and equipment.
- Wide clear pathways and visual and tactile markers.
- Engaging, high-interest learning materials.
- Assistive technology is available and used.

Language and communication access

- Total communication approach is often best approach.
- Sign language, if used by student, should be primary language used in the class.
- Student's preferred mode of communication is identified and supported.
- Communication skills are taught and practiced in EVERY lesson, activity, and routines.
- Communication partners are attentive and responsive.
- Classmates and peers receive information about how the student communicates and reciprocal interactions are supported.

Meaningful curriculum

- Use the child's interests to engage them.
- Academic materials and lessons may need modifications.
- Modify to accommodate vision, hearing, motor, and balance issues.
- Provide multiple ways for student to demonstrate what they know and what they've learned.
- "Curriculum" includes learning new routines, self-care skills, social skills, independent living skills, problem-solving and goal setting.

Sensory needs are met

This is a critical area to address!

- Student may need frequent breaks to rest or to move.
- Posture, position and movements the child uses or engages in serve a purpose. Identify purpose and provide these opportunities to student.
- Activities and materials that offer sensory input student is seeking are provided in a consistent and predictable way.
- Activities and materials that the student avoids are not present or limited.
- Sensory needs may change in different environments.

Social relationships

Everyone seeks connections. Students who are still learning to communicate effectively seek connection through "behaviors".

- Adults must share inform and support interactions and shared activities between child and classmate or peers.
- Create opportunities for turn-taking and sharing within activities.
- Allow peers to model and support child when possible.
- Point out and practice social skills the child may not have learned incidentally.
- Connect students through common interests.

Proper Support

Proper support will help to ensure learning goals and the previous six elements can be implemented. Proper support services will…

- Expand communication and concept development.
- Support the child in maintaining sensory regulation and managing behavior.
- Notice and attend to any medical and health needs and manage assistive technology.
- Ensure environment and materials are accessible.
- Promote independence and self-determination.

References

- Brown, D. (2014). What does "Follow the Child" mean?. *reSources, 19* (1). 1-8.E
- Brown, D. (2001). Follow the child: Approaches to assessing the functional vision and hearing of children with congenital deafblindness. *reSources, 10* (9). 1-3.
- Slavin, L. J., (2018). The educational checklist for individuals with CHARGE syndrome. *https://www.chargesyndrome.org/wp-content/uploads/2020/09/CHARGE_Educational_Checklist_Final.pdf*.
- Nelson, C., van Dijk, J., McDonnell, A., & Thompson, K. (2002). A framework for understanding young children with severe disabilities: The van Dijk approach to assessment. Research and Practice for Persons with Severe Disabilities, 27, 97-111.