

Making school days some of the best days of their lives for young people with single ventricle conditions

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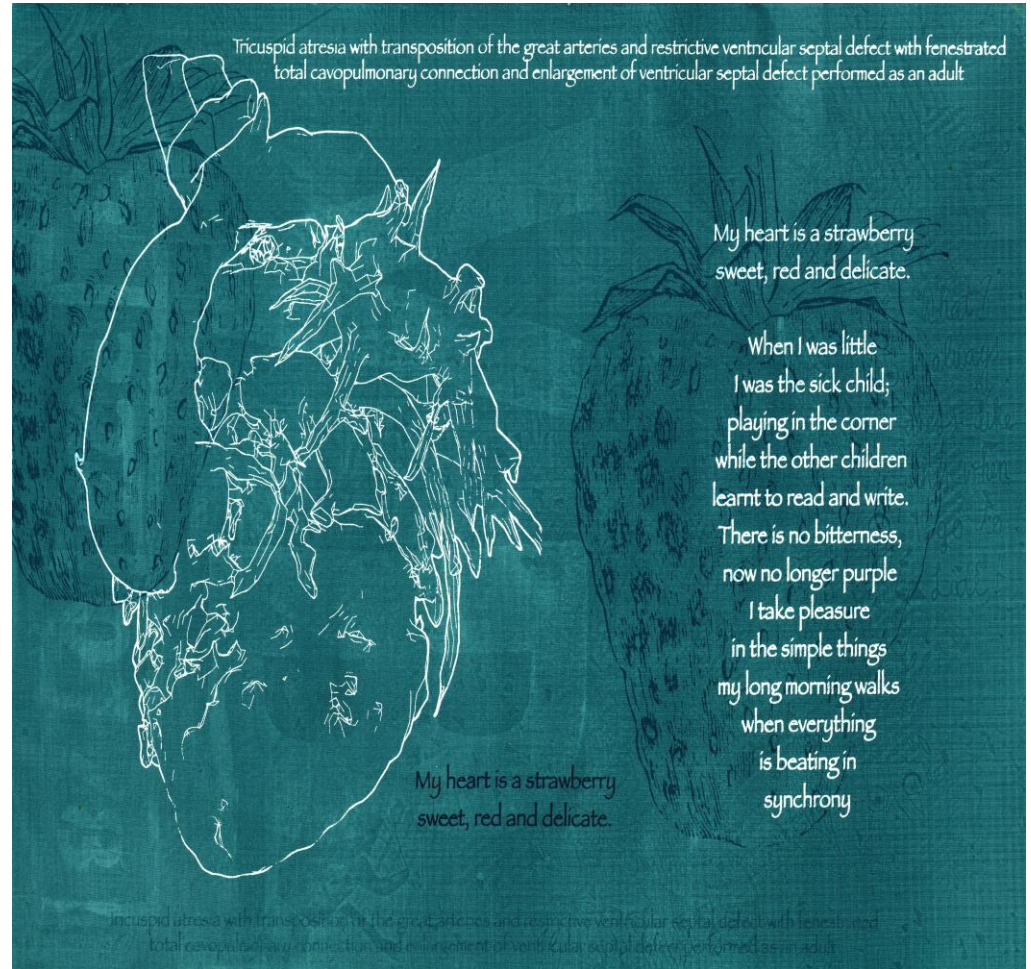
SLIDO question



As increasing numbers of children with complex CHD require educational support against a backdrop of diminishing resources, what is our best contribution as health professionals?

Adult SV patient

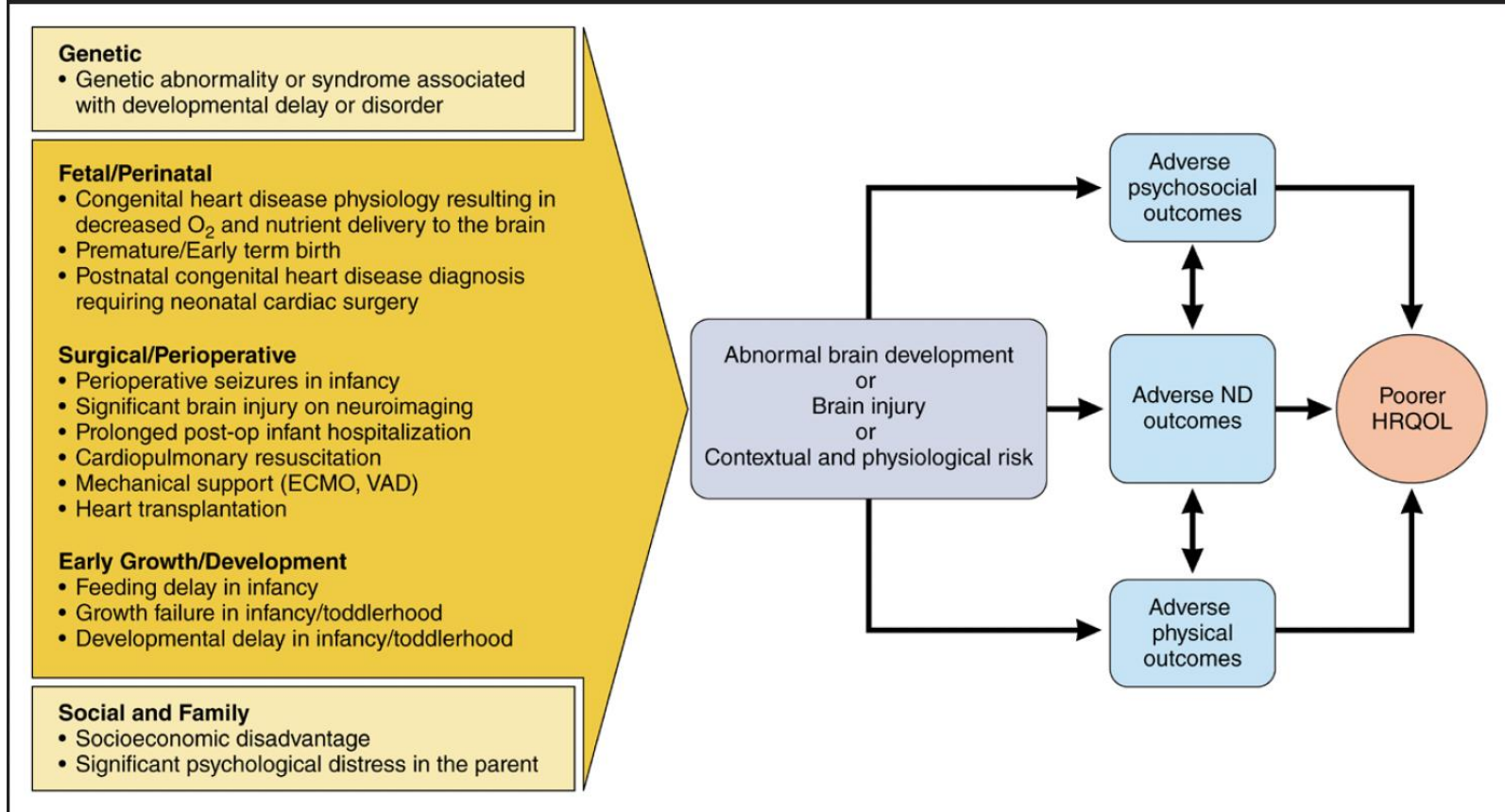
- Not taught to read or write at school because of CHD
- Fatigue – excluded from all activities
- Prevented from living their best life



Tasks of childhood and adolescence

- **Physical development** – participation in activities, development of more complex motor skills, strength, balance and coordination,
- **Social development** – increasing independence, problem solving, leadership opportunities, cooperation, functioning individually and as part of a team, shared expectations, peer relationships, separation from family
- **Cognitive development** – reasoning skills, leadership skills, decision-making and problem solving, development of identity
- **Emotional development** – success in meeting challenges, feelings of self-worth, mood swings, experiencing success, use of role models
- All of which can be significantly impacted by CHD – **delays** to normal developmental trajectory, some aspects **not achievable**, **disruptions** due to illness/routine medical follow-up... etc and all of which happens in a network of systems

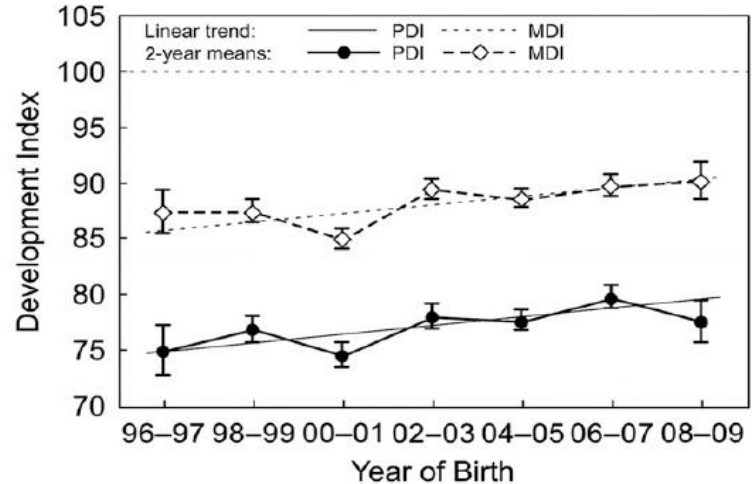
Factors that increase neurodevelopmental risk



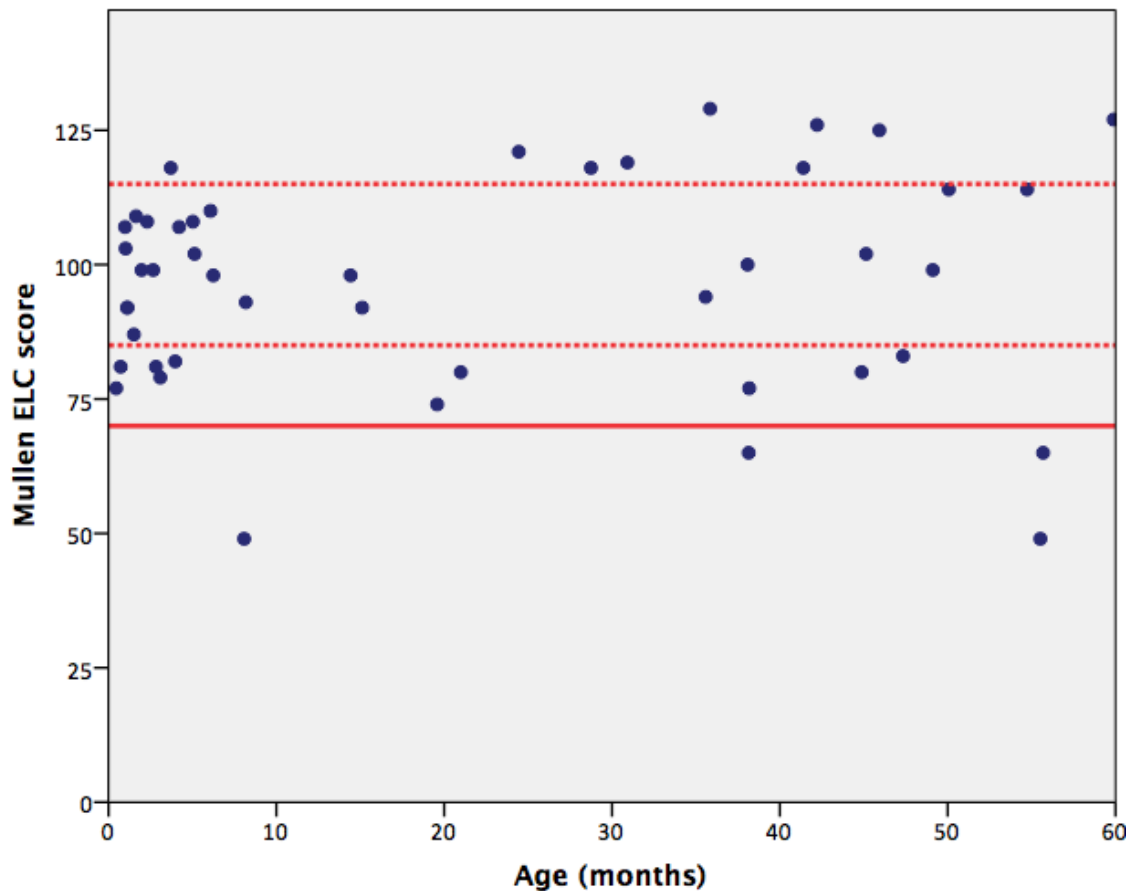
Neurodevelopmental Outcomes After Cardiac Surgery in Infancy

- 1770 subjects
- 22 centers
- BSID-II at 14.5 months
- More children with more complex CHD in later years

- For whole cohort – early ND outcomes showed modest improvements over time after adjusting for innate patient risk factors (lower birth weight, male gender, less maternal education, presence of suspected or definite genetic diagnosis)
- **No improvements for HLHS patients after adjustment for patient factors**
- As more high-risk infants with CHD survive surgery, increasing numbers will require significant societal resources



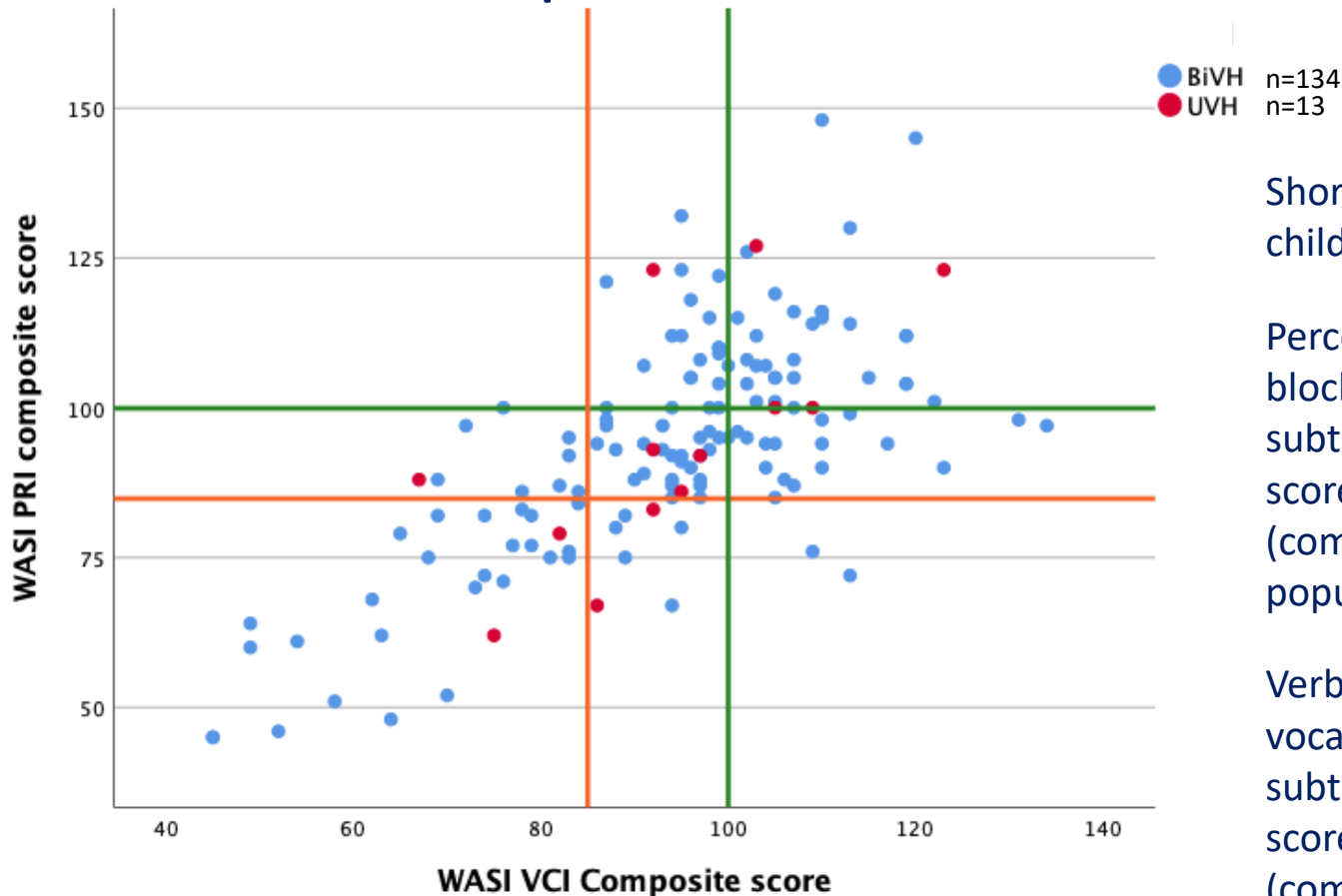
Pre-school: Mullen ELC (early learning composite – fine motor, visual reception, expressive and receptive language skills): total scores (SV: n=46)



Cognitive outcomes at school age

- IQ in normal range but at the lower end of normal; increased rate of lower IQ scores
- Visual motor skills, working memory, processing speed and executive functioning - areas of particular delay
- Increased incidence of attention problems; up to two-thirds show evidence of ADHD
- Increased incidence of ASD
- Significant proportion score in 'at-risk' range on multiple domains of ND functioning
- Higher incidence of academic problems and requirement for educational support
- Deficits continue into adulthood

IQ data – UK sample

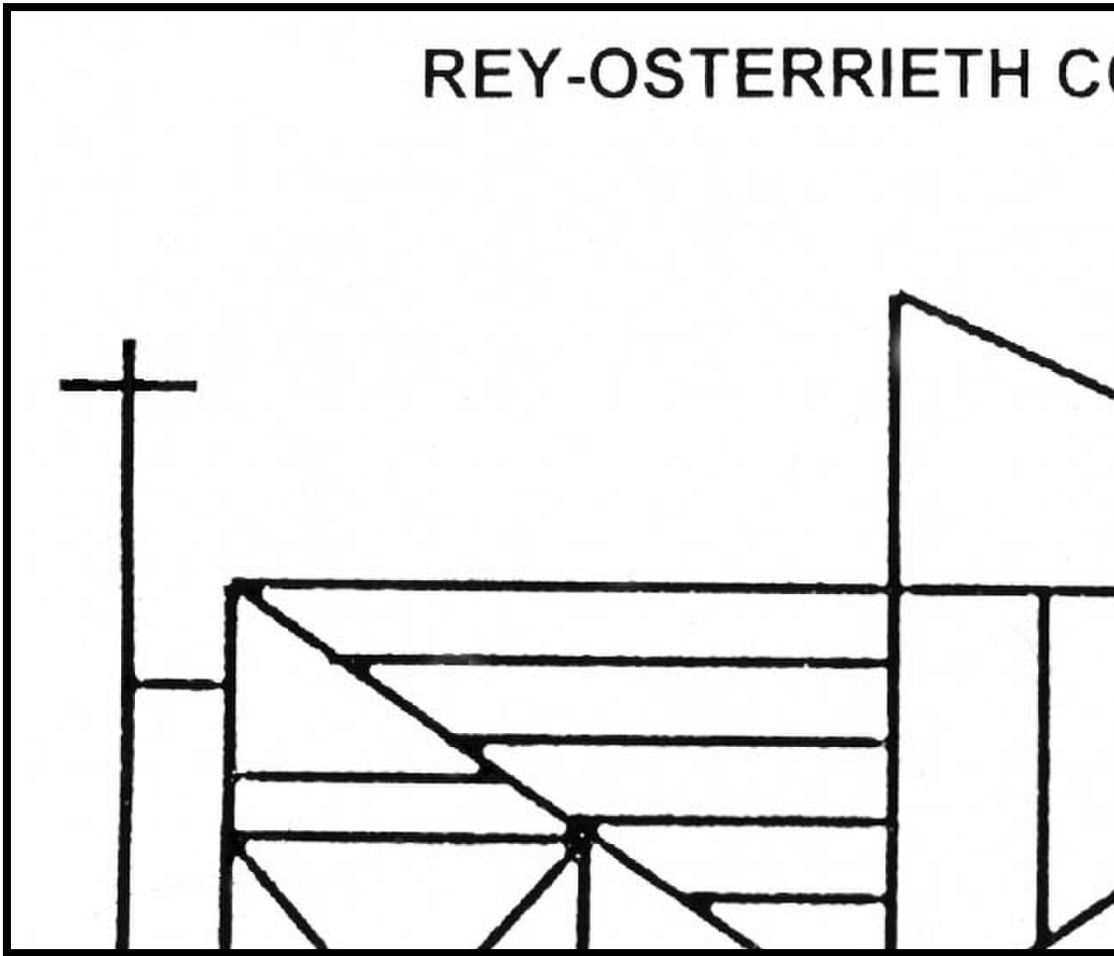


Short form IQ test (WASI) – 164 children aged 5-16 years

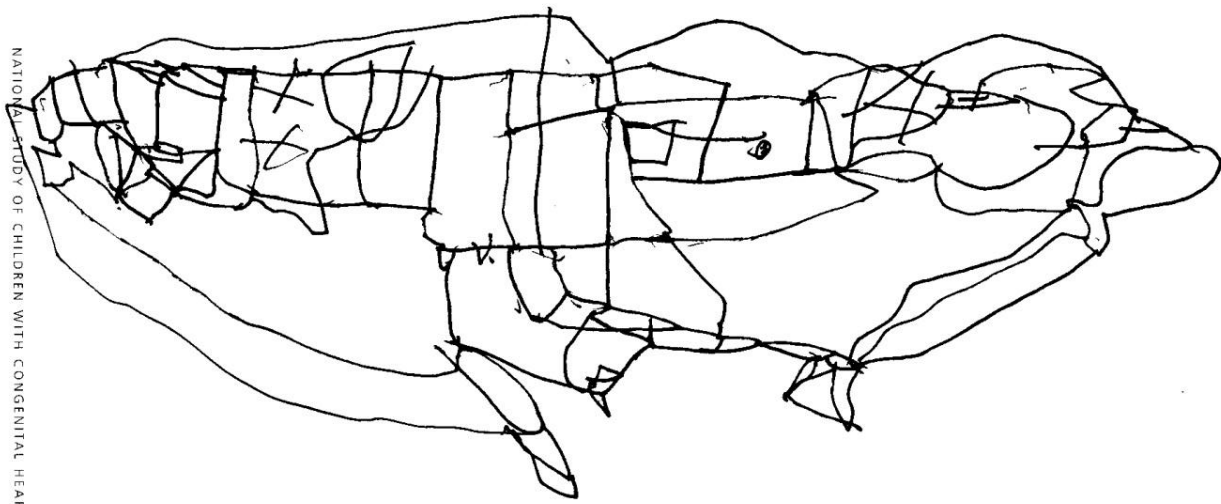
Perceptual reasoning index (PRI – block design and matrix reasoning subtests): 31% of SV patients scored >1SD below mean (compared with 16% in healthy population)

Verbal intelligence index (VCI – vocabulary and similarities subtests): 15% of SV patients scored >1SD below mean (compared with 16% in healthy population)

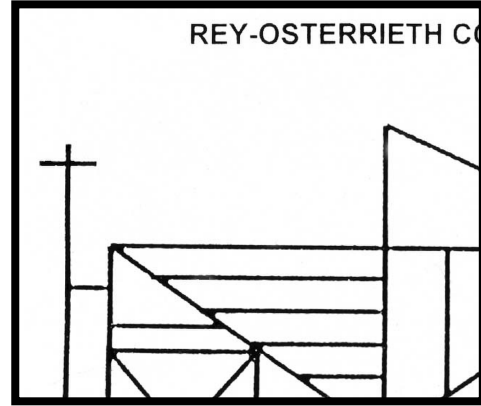
REY-OSTERRIETH CO



Assesses range of neuropsychological functions – visuospatial abilities, attention and planning are assessed in the copying (recognition) task

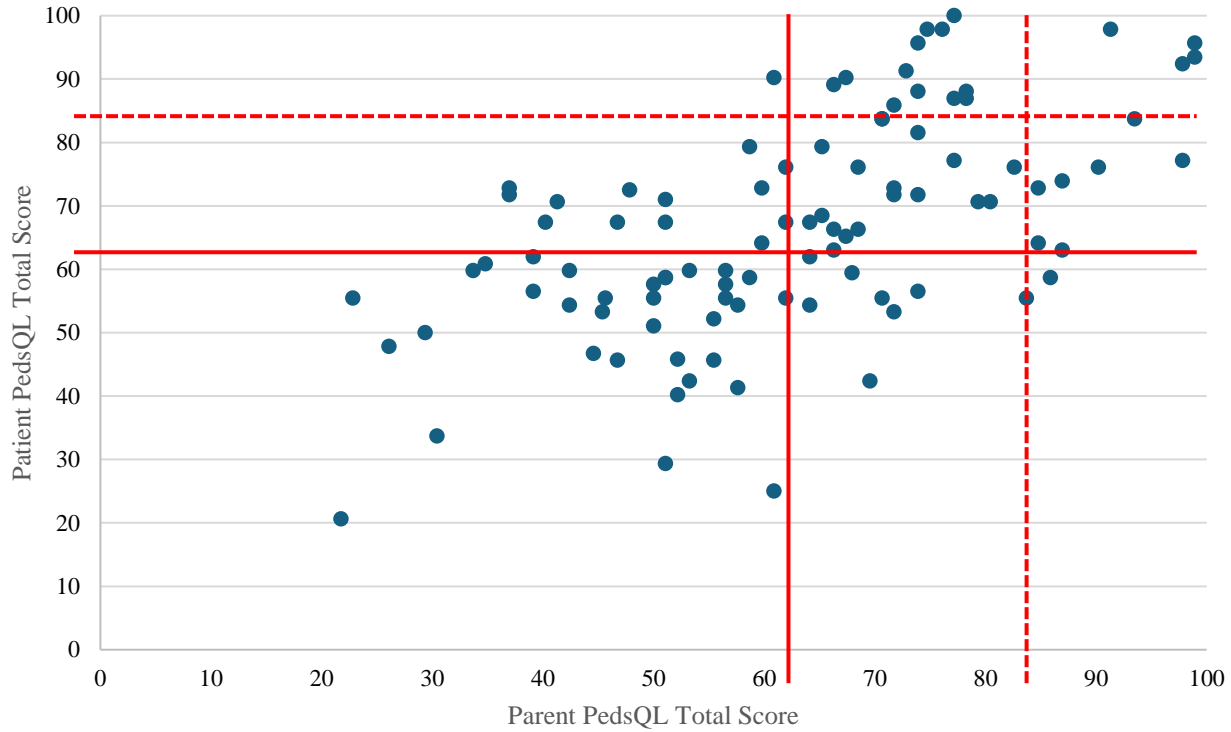


Boy, 10.5 years



With thanks to Dr Rachel Knowles for permission to use the image

Child and parent PedsQL scores – generic QoL measure



N=99

Correlation between child and parent rating= 0.59

Healthy means

Parent total: 85

Child total: 84

What do we need?

- Information – lots out there (LHM!) – how often do we signpost to these?
- Consistent use of individual and education health care plans
- Guidelines – nothing is routinely implemented in the UK
- Screening tools – development, mental health etc
- Tools for monitoring – e.g. PROMs
- Tools to help young people communicate
- School-based interventions
- Resourced pathways for screening/evaluation/monitoring/intervention
- What else...?



Support for a Child with Special Educational Needs (SEN) within Nursery, School or Further Education. Little Hearts Matter



Understanding what having half a working heart means for a child in school
A guide for primary and secondary school teachers

Developmental Screen – 15 months–2;11 years (score: Yes/No) Insert Patient sticker here

Child's age: ____ Years ____ Months Date of assessment: __/__/__

1. Gross motor skills (score 0-4) Achieved Age (months)

Walks alone	15-20	0
Can jump off step	21-25	0
Can stand on one foot (5+ seconds)	26-30	0
Walks upstairs, one foot on each step, adult manner	31-35	0
Total	3;4	0

2. Fine motor skills (score 0-4) Achieved Age (months)

Builds a tower of 4 blocks	15-20	0
Makes a brick or toy walk	21-25	0
Copies a line	26-30	0
Copies a cross (imagetable)	31-35	0
Total	2;4	0

3. Daily living skills (score 0-4) Achieved Age (months)

Small clothing	15-20	0
Indicates toilet needs	21-25	0
Helps put toys away	26-30	0
Dresses and undresses self except for buttons	31-35	0
Total	2;4	0

4. Communication (score 0-4) Achieved Age (months)

Uses 7+ clear words	15-20	0
Narrates 4 objects	21-25	0
Defines by use 2 objects (eg. knife, coat, car)	26-30	0
Uses 2 personal pronouns	31-35	0
Total	2;4	0

5. Socialisation (score 0-4) Achieved Age (months)

Shows desire to please others	15-20	0
Plays alongside others – parallel play	21-25	0
Gives name on request	26-30	0
Plays with other children	31-35	0
Total	2;4	0

6. General understanding (score 0-4) Achieved Age (months)

Knows 3+ body parts	15-20	0
Listens to stories with pictures	21-25	0
Compares 2 objects – which one is bigger	26-30	0
Knows age and sex	31-35	0
Total	3;4	0

Gross motor score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

Fine motor score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

Daily living score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

Communication score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

Socialisation score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

General understanding score
1;4 0 0
2;4 1 0
3;4 2 1
4;3 3 5/2

Do the parents have any concerns about their child?
Speech; does not get on well with other children

Languages spoken at home English
Child's primary language English

Pediatric Cardiac Quality of Life Instrument (PCQLI) Adolescent Form

	Excellent	Very Good	Good	Fair	Poor
1 In general would you say your health is ...	1	2	3	4	5
Because of my heart problem...					
2 I feel different from everybody in a bad way	1	2	3	4	5
3 I can't do the physical activities I want to do	1	2	3	4	5
4 I miss too much school/college/work	1	2	3	4	5
5 I feel guilty about the stress my heart disease causes my family	1	2	3	4	5
6 School/college work is difficult for me	1	2	3	4	5
7 I get unwanted attention	1	2	3	4	5
8 I am afraid of medical procedures	1	2	3	4	5
9 I get tired easily	1	2	3	4	5
10 I take too much medicine	1	2	3	4	5
11 Adults around me are overprotective	1	2	3	4	5
12 I have no energy	1	2	3	4	5
13 I hang back when I am doing physical activities	1	2	3	4	5
14 Other people are uncomfortable around me	1	2	3	4	5
15 I am in pain	1	2	3	4	5
16 I am busy to have other health problems	1	2	3	4	5
17 I cannot wear what I want	1	2	3	4	5
18 I spend too much time dealing with my health	1	2	3	4	5
19 I take medicine that causes bad side effects	1	2	3	4	5
20 My heart condition is likely to get worse	1	2	3	4	5
21 I get special treatment at home/school/work	1	2	3	4	5
22 I miss social activities	1	2	3	4	5
23 I engage in risk taking behaviours	1	2	3	4	5
24 I am afraid of dying	1	2	3	4	5
25 It is difficult to get around from place to place	1	2	3	4	5
26 I feel angry	1	2	3	4	5
27 Other people treat me differently	1	2	3	4	5
28 I look different from everybody in a bad way	1	2	3	4	5
29 I worry about my future	1	2	3	4	5
30 I feel helpless	1	2	3	4	5

IHCPs and EHCPs

- Individual healthcare plans (IHCPs) are specifically for children with medical needs:
 - Set out child's medical condition, any support needed and any emergency procedures
 - No standard national format for an IHCP; schools can draw up their own with contributions from parents, health professionals etc.
 - **All children with SV conditions are eligible for an IHCP**
- Education, health and care plans (EHCPs) are education driven and are for children/young people (until 25 years old) with special educational needs:
 - Children who have significantly greater difficulty in learning than others of their age and/or children with a disability who can't access educational facilities normally available in mainstream school
 - Assessment is carried out by local authority to determine if a child needs an EHCP and what support should be in it
 - Majority if not all children with SV conditions are eligible for an EHCP
- Children can have **both** an IHCP **and** an EHCP



Neurodevelopmental Outcomes in Children With Congenital Heart Disease: Evaluation and Management

A Scientific Statement From the American Heart Association
 This statement has been approved by the American Academy of Pediatrics.

Bradley S. Marino, MD, MPP, MSCE, FAHA, Co-Chair; Paul H. Lipkin, MD; Jane W. Newburger, MD, MPH, FAHA; Georgina Peacock, MD, MPH; Marsha Gerdes, PhD; J. William Gaynor, MD; Kathleen A. Mussatto, PhD, RN; Karen Uzark, PhD, CNP, FAHA; Caren S. Goldberg, MD, MS; Walter H. Johnson, Jr, MD; Jennifer Li, MD; Sabrina E. Smith, MD, PhD; David C. Bellinger, PhD; William T. Mahle, MD, FAHA, Co-Chair, on behalf of the American Heart Association Congenital Heart Defects Committee of the Council on Cardiovascular Disease in the Young, Council on Cardiovascular Nursing, and Stroke Council

2012



Guidelines for neurodevelopmental follow-up

Neurodevelopmental Follow-up of Children with Congenital Heart Disease in Europe

Johanna Calderon (France), Bettina Reich (Munich), Andrew Chew (London), Monique van Schooneveld (Utrecht), Jo Viray (London), Frank Casey (Belfast) Pascal Amedro (Bordeaux), Damien Bonnet (Paris), Katya de Groote (Gent), Bea Latal (Zurich) and the EU-ABC Europe Working Group* and the AEPC, supported by the CNOC
 January 2023

* On behalf of each participating EU cardiac neurodevelopmental teams

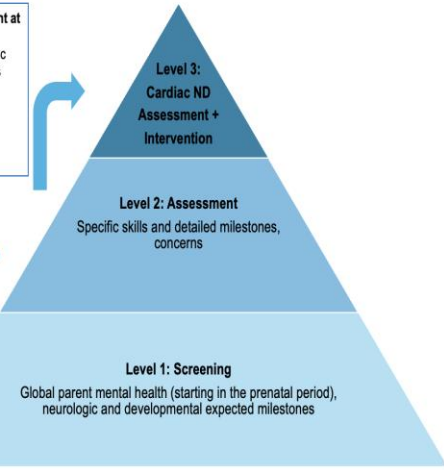


CHD groups for follow-up : Individualized Risk Stratification Model

Criteria for expert diagnosis and treatment at for highest neuro risk:
 Diagnosis of CHD requiring **neonatal** cardiac surgery (i.e., HLHS), or severe complications ECMO, VAD, prematurity, clinical seizures, abnormalities in MRI, or based on clinical judgment AND/OR
 -Positive ND concerns at the Assessment Level

Positive ND screening Level 1 and/or any of the criteria for expert ND diagnosis & treatment

- ALL diagnoses of CHD requiring cardiac surgery up to 12 mo
 -Any CHD, any intervention timing with ND concerns
 - Any CHD diagnosis with significant comorbidities as defined by provider



Marino et al, 2014; Sood et al, 2024; Calderon et al, 2024

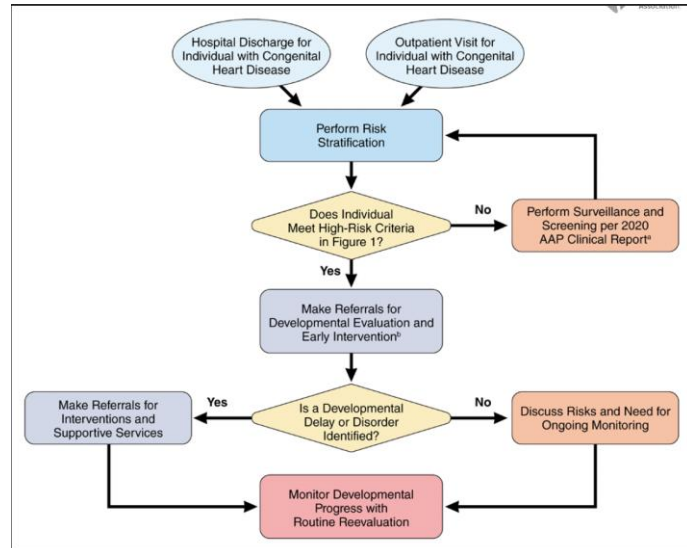
AHA Scientific Statement

Neurodevelopmental Outcomes for Individuals With Congenital Heart Disease: Updates in Neuroprotection, Risk-Stratification, Evaluation, and Management: A Scientific Statement From the American Heart Association

Endorsed by the Cardiac Neurodevelopmental Outcome Collaborative

Erica Sood, PhD, Vice Chair; Jane W. Newburger, MD, MPH, FAHA; Julia S. Anixt, MD; Adam R. Cassidy, PhD, ABPP; Jamie L. Jackson, PhD; Richard A. Jonas, MD; Amy J. Lisanti, PhD, RN, CCNS, FAHA; Kella N. Lopes, MD, MPH; Shabnam Peyvand, MD, MAS, FAHA; Bradley S. Marino, MD, MPP, MSCE, MBA, FAHA, Chair, on behalf of the American Heart Association Council on Lifelong Congenital Heart Disease and Heart Health in the Young and the Council on Cardiovascular and Stroke Nursing

2024



Algorithm for referral, evaluation and management for individuals at high risk (Sood et al, 2024)

The challenge for the UK and Europe...



- European survey
- Only 2 UK centres participated, one in London, one ?level 2 centre
- 10/25 had ND follow-up programme
- 6/25 planning to implement ND follow-up programme
- But were any of these UK centres....?
- Informal survey of UK SIGs in cardiac psychology – very little in place...
- Resources – significant issue

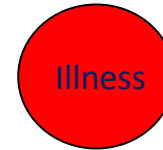
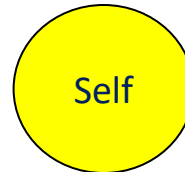
PRISM – Pictorial
Representation of
Illness and Self
Measure – a
measure of illness
burden

(Buchi et al, 2002)

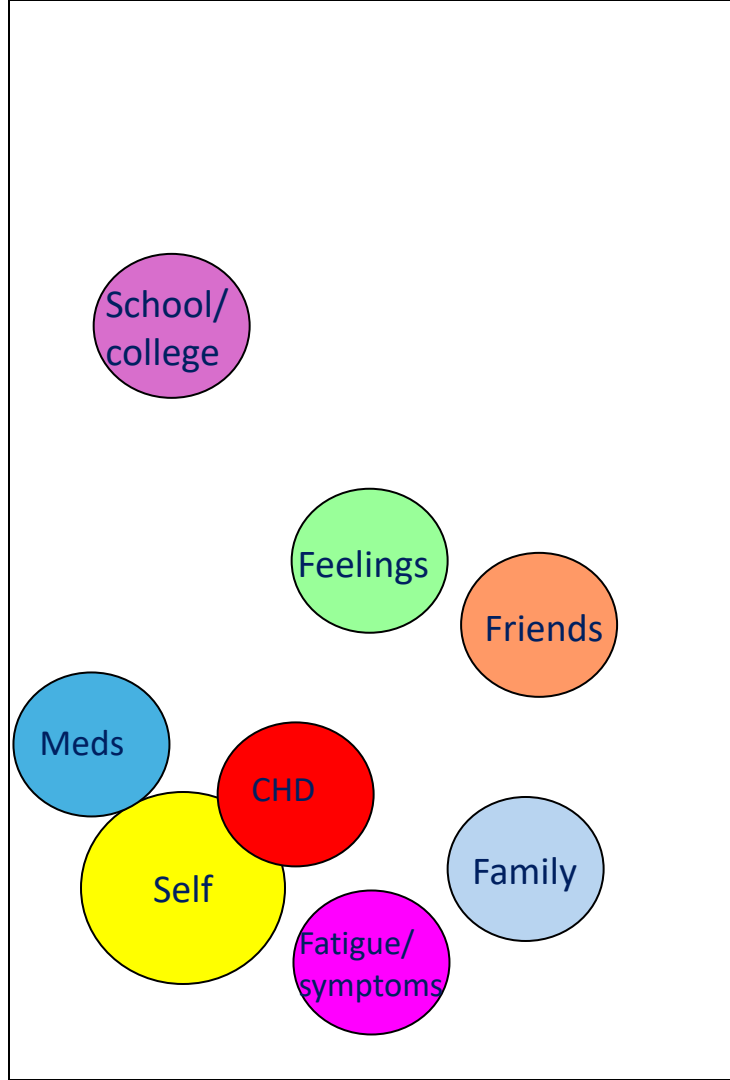
This white sheet represents
your life as it is now.....

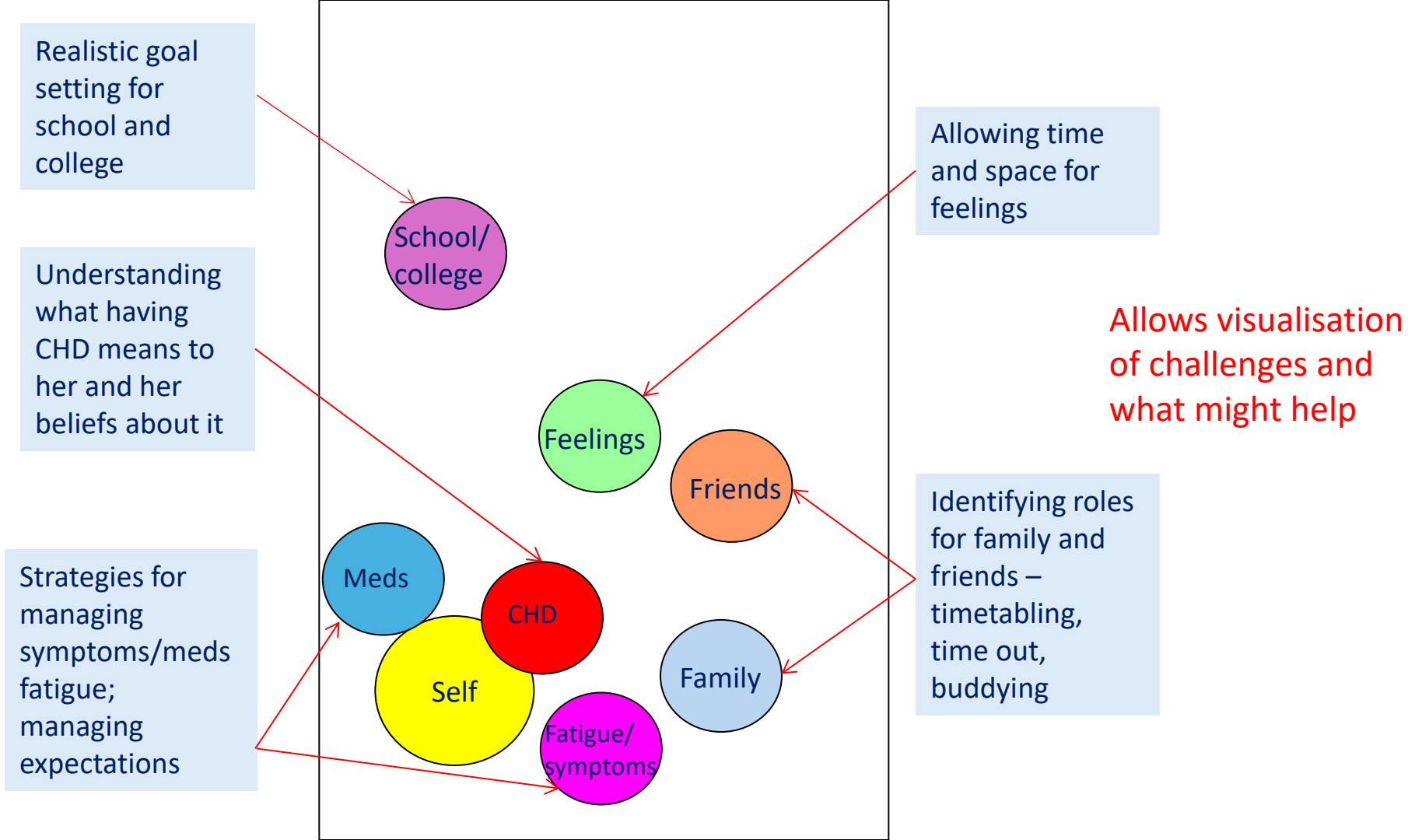
The yellow disk represents
your “self”

And this red disc represents your
illness. Where would you place
your illness – the red disc – in
your life at the moment?



Chloe





Spoon theory - a metaphor describing the amount of physical or mental energy that a person has available for daily activities and tasks, and how it can become limited (Miserandino 2003)

Can help to make an individual 'spoon map'

With only 10 spoons of energy it doesn't take long before the spoons need to be replenished... and some days it might be less than 10 spoons



get out of bed



bathe



eat a meal



go to work/school



get dressed



brush hair



make plans & socialize



go shopping



take pills



surf the internet



homework



go to the doctor



watch TV



read a book



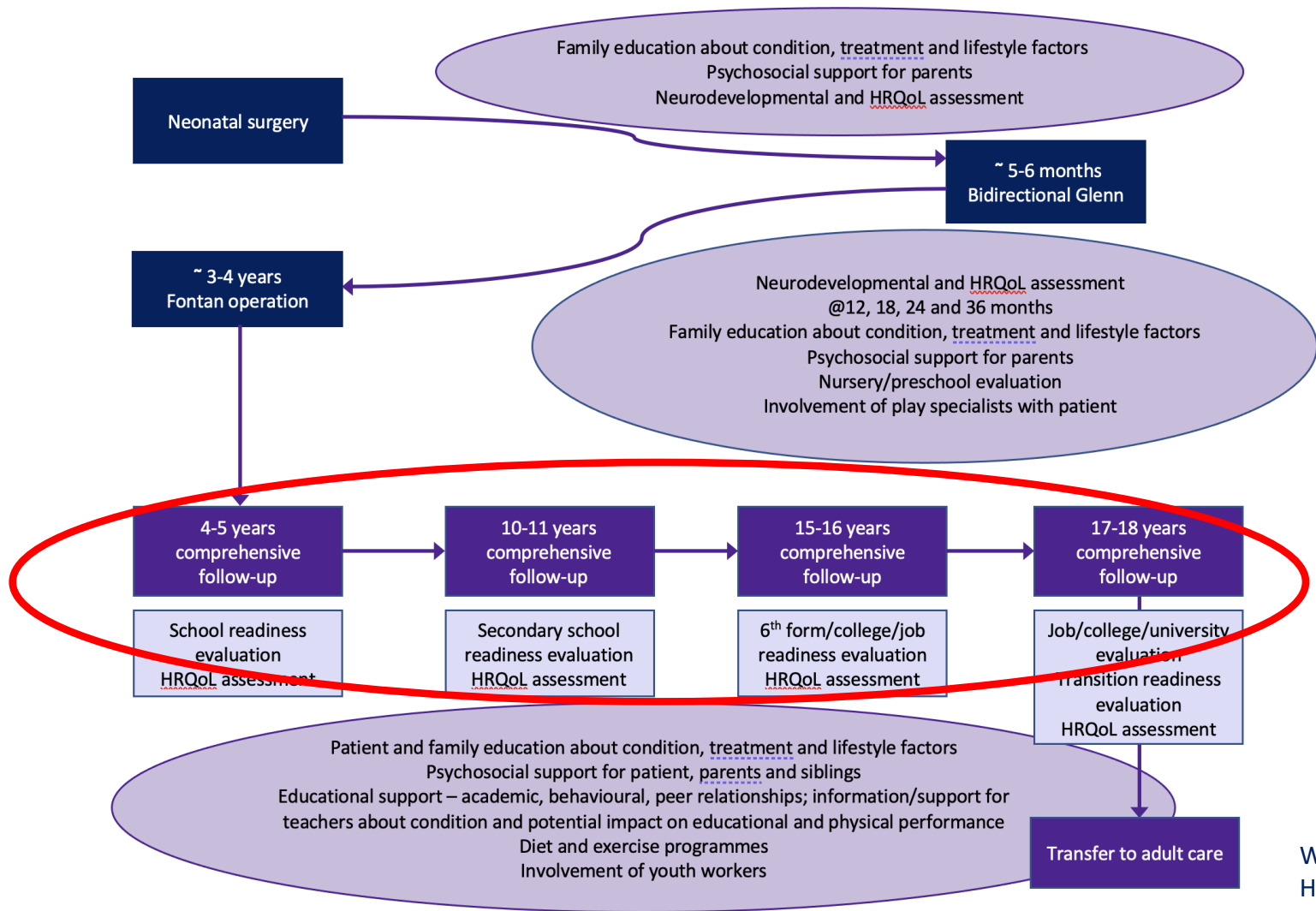
drive somewhere



exercise


Examples of school-based interventions for children with CHD

- Education of teachers, educational psychologists, teaching assistants and administrators about the child's medical condition, the risk for developmental delays and specific learning needs
- Avoidance of diagnostic overshadowing/labelling
- Routine neuropsychologic or psychoeducational evaluations
- Providing extended time for homework, exams, and projects
- Providing “note taker” and/or recorded classes
- Assistance of a reader or scribe for exams
- Quiet room with decreased distractions for exams
- Physical, occupational and speech therapies
- Keyboarding as an option instead of handwriting



We can do this better...

- MDT with health and education professionals
- Attendance at review meeting by psychologist; school visit by psychologist prior to starting secondary school
- Ensuring all teachers informed; communication plan
- Early conversations about what comes after school
- Fatigue – significant problem
 - Timetable reduced
 - Rescheduling of lessons/classrooms to reduce distance to walk; access to lift
 - Shorter school day
 - Note takers/extra time/organisational planner
 - Regular snacks
 - Played goalkeeper for the best team at school
- Support for psychological issues related to fear of dying

A large, white iceberg floats on the surface of the ocean under a blue sky with scattered clouds. The water is a clear, vibrant blue. Below the surface, a scuba diver is visible, swimming horizontally. The iceberg's surface is textured with shadows and highlights, suggesting its massive scale. The diver is positioned in the lower right quadrant of the frame, providing a sense of scale to the iceberg.

If we can get it right for our most complex patients we can get it right for all...

Thank you!