
Learning and behaviour in Noonan syndrome

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Overview

What we will cover today

1

What the research shows

Learning, behaviour and wellbeing at each stage of growing up



Preschool

Development, movement, sleep



Primary school

Learning, ADHD, autism, anxiety



Adolescence

Feelings and independence

2

How and where to ask for help

General principles for managing learning and behaviour, and what to do while you wait

3

Why we need more research

Our ongoing studies, and how understanding the brain could lead to better treatments

Every person with Noonan syndrome is different

1

Research describes groups groups

Numbers in this talk are averages
averages from studies. They cannot
cannot predict how any one person
person will learn or behave.

2

The range is wide

Many children and adults learn and
and get on well. Others need extra
extra help at school, at work or at
at home.

3

Knowing helps

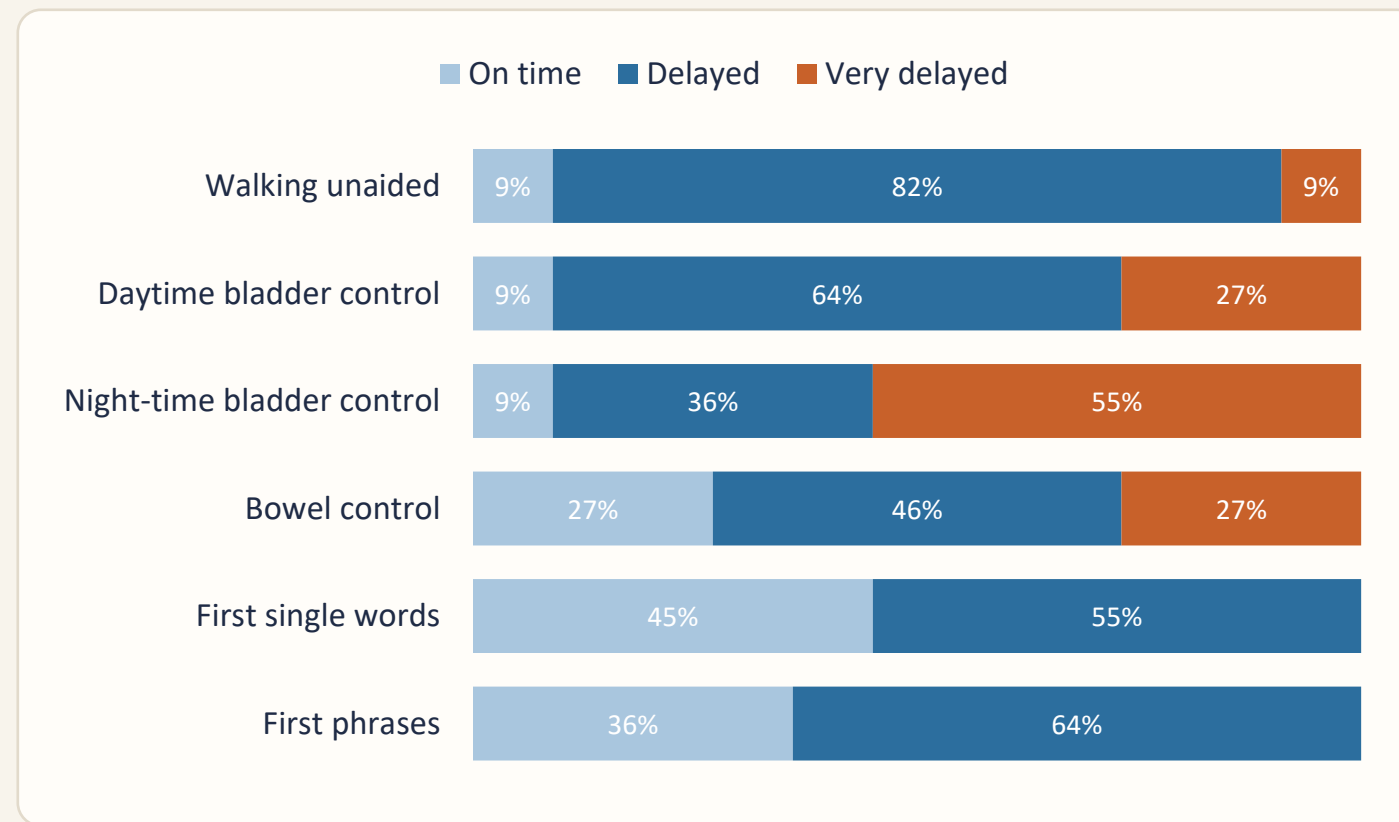
Knowing what can happen helps you
you spot things early and ask for the
the right support.

Noonan syndrome: learning and behaviour from infancy to adolescence



Infancy and pre school period

Early milestones in children with Noonan syndrome



Most walk a little later

About 8 in 10 were somewhat delayed; few were very delayed



Toilet training takes longer

Especially staying dry at night



Talking comes on steadily

Nearly half said first words on time; none were very delayed

Infancy and pre school period- Sleep



Commonly reported by parents in clinic

Clinically, one of the things families raise most often



Not well studied in infancy

Very little research in babies and toddlers so far



Sleep is vital in early development

It helps the developing brain with learning and memory

PRIMARY SCHOOL AGE

1 in 4



children with Noonan syndrome have sleep difficulties (about 25%)



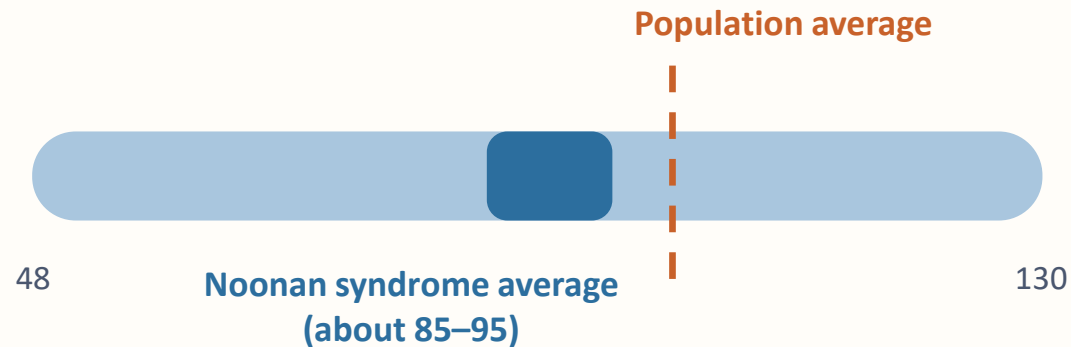
Most common in clinic

Difficulty falling asleep

School age: learning

Most children with Noonan syndrome learn within or close to the typical range

THE RANGE OF ABILITY IS WIDE



Individual scores in one study ranged from 48 to 130

About 1 in 4

have an intellectual disability



Variant make a difference

SOS1: usually average or above.
PTPN11, KRAS, RAF1 and SHOC2
and other variants are more
variable

Attention and working memory are the main challenges

Working memory

Holding information in mind while using it, such as following a set of instructions.

Attention

Staying focused, especially on long tasks or ones that feel less interesting.

Processing speed

Needing a little longer to take things in and respond. This can continue into adulthood.

These differences remain even after taking overall ability into account, so they are worth supporting in their own right.

School age :Autism/ neurodiversity

Rates of autism are between 10–30%, compared with 1–2% in general population

OUR MANCHESTER STUDY (CASPER): 40 CHILDREN, DETAILED ASSESSMENT

30%

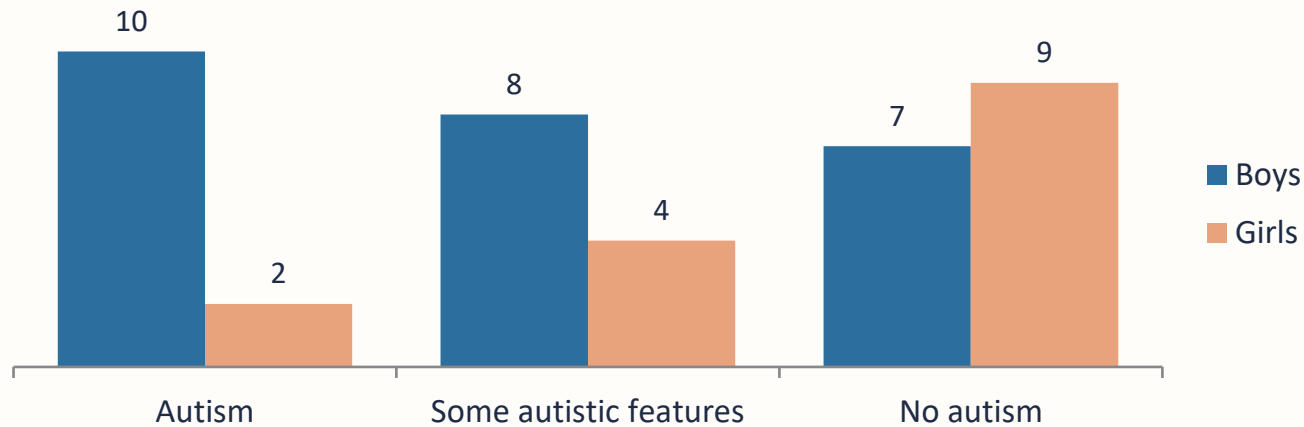
Autism

30%

Some autistic features

40%

No autism



5 boys to 1 girl
among children with autism



52% also had ADHD
IQ and ADHD were similar
whether or not a child had autism

School age: autism/ neurodiversity

WHAT IT CAN LOOK LIKE



Making and keeping friends



Coping with change



Sensitivity to noise, touch or textures

LINKED TO THINGS



Attention

Fewer ADHD symptoms means stronger social skills



Irritability and emotions

Strongly tied to social difficulty in Noonan syndrome



Does the gene matter?

Autism-related traits in 128 people, by gene

PTPN11

Most autism-related difficulties; stronger gene effect linked to more repetitive behaviours

SOS1

Also more autism-related traits than other children

RAF1

Fewer autism-related difficulties than PTPN11

School age: ADHD

About half of children meet criteria for ADHD, compared with around 5% of all children

About 1 in 2

children with Noonan syndrome



About 1 in 20

all children



Up to 7 in 10 children with Noonan syndrome show some signs of ADHD, even without a diagnosis



Attention and working memory

Staying focused, holding instructions in mind



Impulsivity and restlessness

Acting before thinking; finding it hard to wait



Planning and organising

Getting started, keeping track of things

Not explained by IQ: children with and without ADHD had similar overall ability

Movement and coordination

Coordination, strength and stamina are often affected

7 in 10

children scored low on a standard coordination test

6 in 10

parents report pain, weakness, tiredness or clumsiness in daily life

WHAT IT CAN LOOK LIKE



Walking a little later

Early motor milestones are often delayed



Handwriting and fine motor

Linked to how children see shapes and space



Weaker grip and less stamina

Plus more flexible (hypermobile) joints



Aches and pains

Joint pain, and in some, nerve-related pain

School age : Feelings and behaviour

Emotional and behavioural difficulties are more common in Noonan syndrome



Inward: worries and feelings

4 in 10

children have an anxiety disorder in detailed assessments

- Social anxiety in about 1 in 7
- Low mood and emotional ups and downs



Outward: behaviour

1 in 4

children have behaviour difficulties, such as defiance

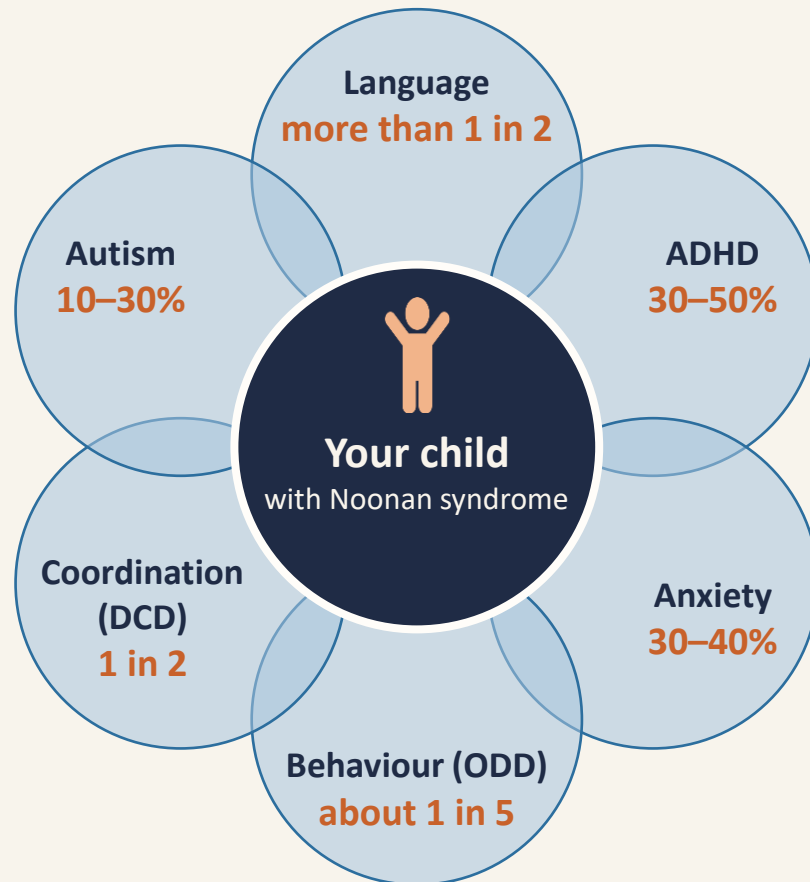
- Frustration, quick temper and aggression
- Often comes alongside ADHD



Irritability links the two. In Noonan syndrome it is closely tied to social difficulties, and these problems are not explained by IQ.

Is it Noonan syndrome or autism or ADHD or just my child?

Often it is several things at once, and each one can be supported



These often overlap

Many children have more than one, so one label rarely explains everything



Look at the whole child

An assessment that considers all of these, not just one, gives the clearest picture



Labels open doors

Noonan syndrome is the cause; a diagnosis like ADHD or autism unlocks support



Each can be helped

Speech therapy, OT, ADHD treatment and support for anxiety all work

Managing learning and behaviour

General principles: the same treatments that help other children work in Noonan syndrome too



1. Seek early assessment

- Don't wait: ask if you are worried
- Look at the whole child, not just one label
- Share results with school



2. Medication for ADHD

- We do use stimulant medication, and it works
- Before starting: check the heart, growth, used with GH
- Involve the heart team if there is a heart condition



3. Help with anxiety

- Talking therapies (CBT), adapted to your child
- Parent-led approaches for younger children
- Predictable routines and preparing for change
- Medication can be considered if anxiety is severe



4. Support for parents

- Parenting programmes help with behaviour
- About 1 in 3 parents report high stress: ask for help
- Connect with other families
- Referral to OT, SALT

“I am on a 4-year waiting list for assessment”

WHAT CAN YOU DO?

1

Arm yourself with the evidence

Share it with your GP, paediatrician and school. In Noonan syndrome:

ADHD
about 1 in 2

Autism
10–30%

Anxiety
30–40%

Language
more than 1 in 2

2

Ask for the genetic diagnosis to be taken into account

High rates of ADHD and autism can support a request to prioritise

3

School support

SEN support often needs diagnosis; ask the SENCO, and apply for an EHCP if needed, Parent partnership

4

Strategies at home

Routines, visual timetables, short instructions and parent programmes

5

Keep a record and connect

We need more research

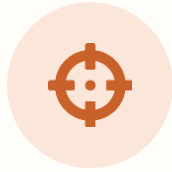
Understanding the brain is the route to more targeted treatments



1. Understand the brain

Brain scans show how Noonan syndrome affects how the brain develops and works

- Brain areas for attention and learning
- How brain cells signal to each other



2. Targeted treatments

Treatments aimed at the cause, not just the symptoms

- Brain stimulation (non-invasive)
- Therapies aimed at the underlying biology



3. Better outcomes

Earlier, more personalised help for learning, attention and wellbeing

- Treatments matched to each person
- Support across the whole lifespan



Coming up next: Izzy, a PhD researcher, will present our brain imaging study

Understanding learning in Noonan Syndrome using neuroimaging



WHAT IS THIS STUDY?

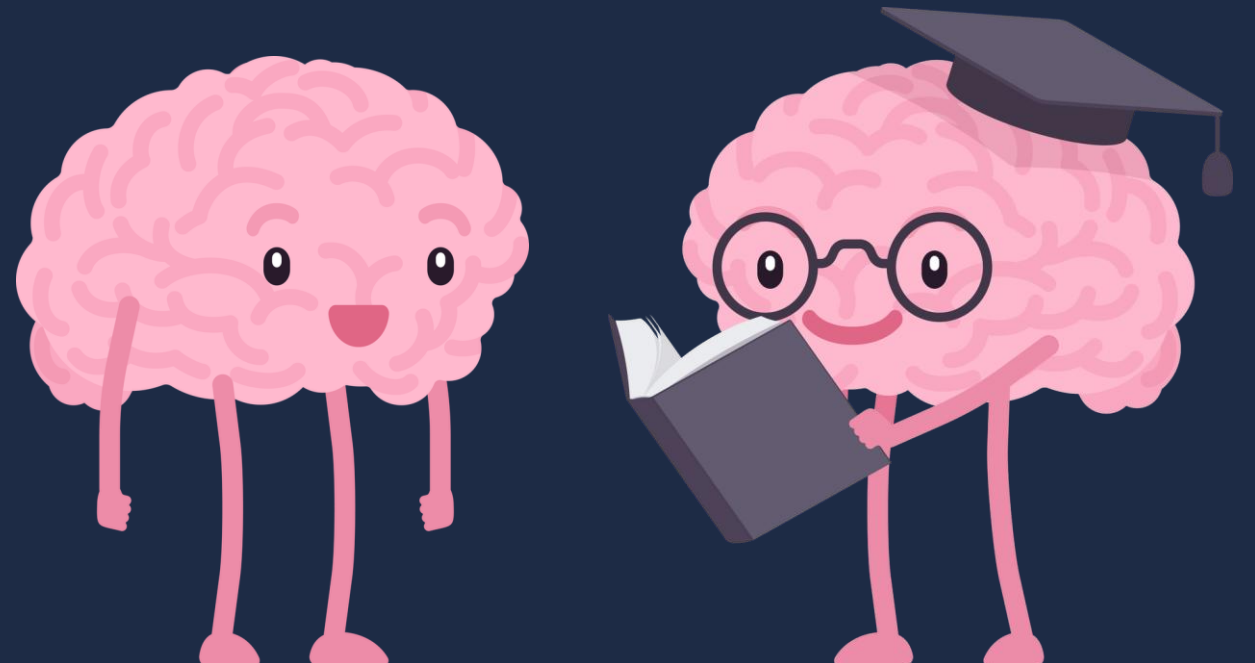
We are recruiting 11 to 17 year olds to participate in a study to learn about Noonan Syndrome and the brain

3-4 hour visit to the research centre

Computerised games

Brain scan

Questionnaires



WHAT DO WE MEASURE?

Interoception

- Trouble regulating emotions
- How do you identify and describe your emotions?
- Can you interpret the state that your body is in?



WHAT DO WE MEASURE?

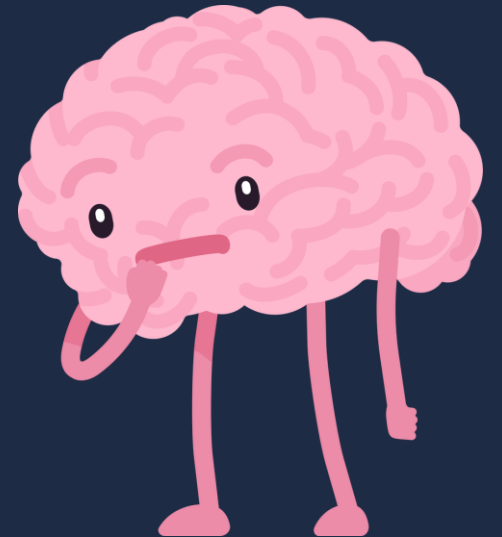
Learning

Working Memory

- Holding information in mind while using it, e.g., following instructions

Long-Term Memory

- Identifying or recalling information that has been seen before



WHAT DO WE MEASURE?

Attention and Processing Speed

Reaction Time game, where an individual is required to pay attention in order to spot the trigger on the screen



WHAT DO WE MEASURE?

Emotion and Behaviour

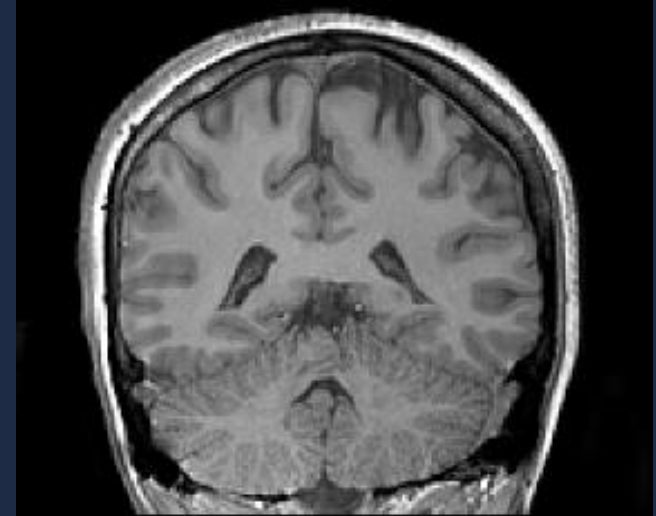
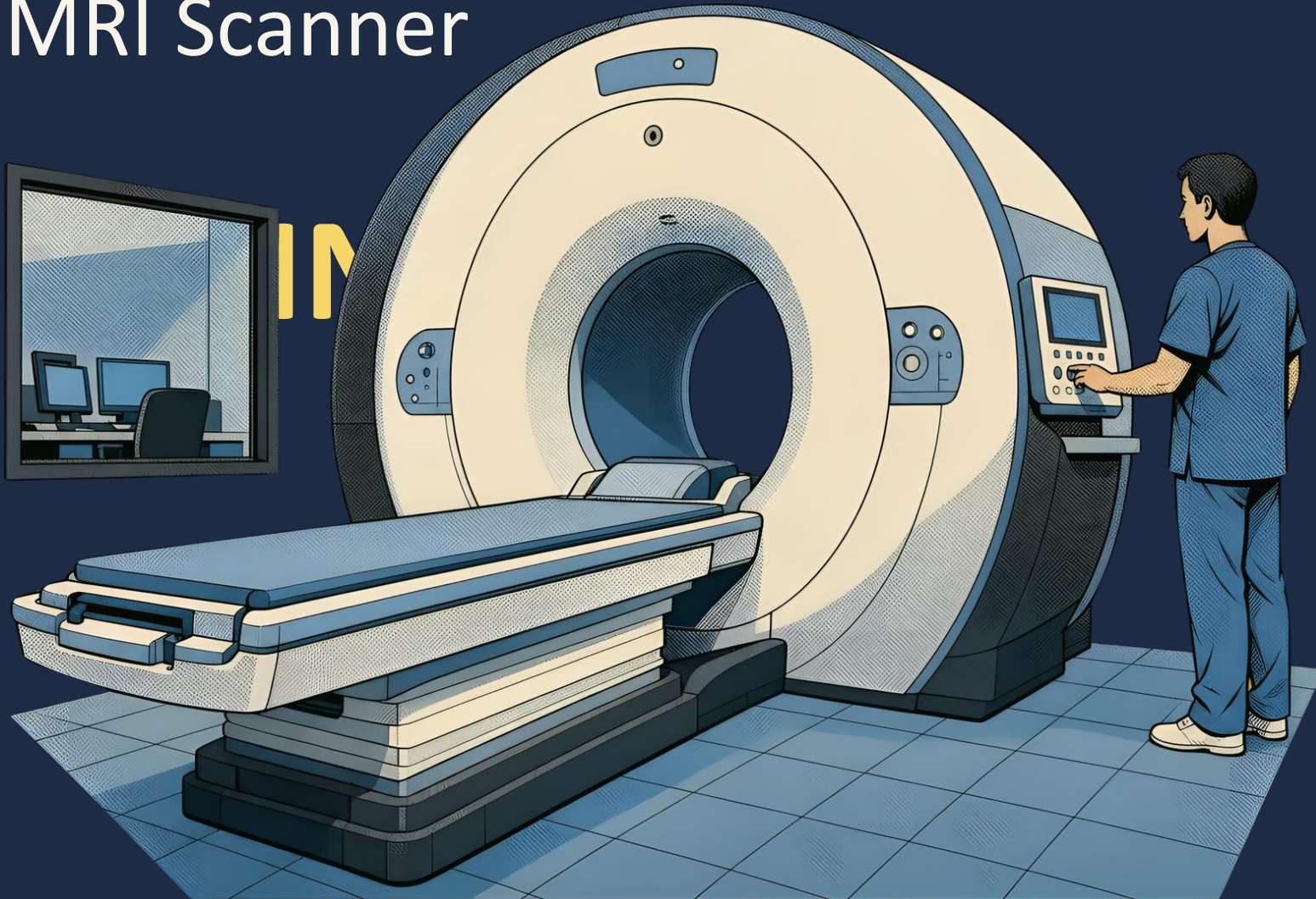
Questionnaires

- Parent and child ratings of wellbeing, anxiety and autism



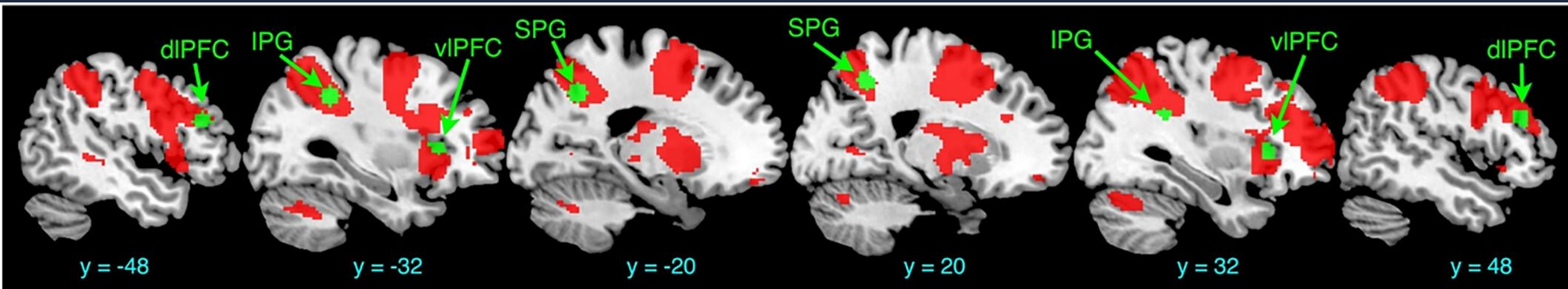
HOW DO WE LOOK AT THE BRAIN?

MRI Scanner



HOW DO WE LOOK AT THE BRAIN?

MRI Scanner



WHAT DO THESE MEASURES TELL US?

Similarities and differences between those with and without Noonan Syndrome

Why some people experience difficulty learning when others do not

How we can help people to cater their learning to their needs



WHAT DO PARTICIPANTS RECEIVE?

- Report of strengths and areas that may benefit from support
- Image of their brain
- Review of brain image by radiology colleagues
- Spending voucher, certificate, stickers, toy
- Travel and food reimbursed in cash



Questions?

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