Socio-emotional functioning of children with Sex Chromosome Trisomies: a focus on emotion regulation

TRIXY

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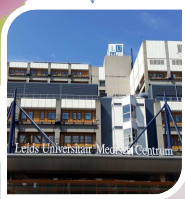
TRIXY Center of Expertise

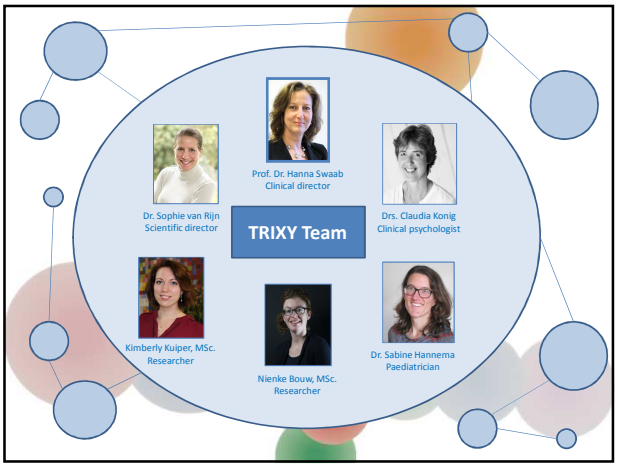
Trisomy of the X and Y Chromosomes

Leiden University Treatment and Expertise Center

Leiden University – Department of Clinical Child and Adolescent Studies



Leiden University Medical Center



TRIXY Center of Expertise

TRIXY Center of Expertise is
the single recognized national Center of Expertise
in the Netherlands for the SCT population
(country with 17.0 million people)

Our objectives:

- Gather scientific knowledge about SCT
- Improve clinical care

The TRIXY Study

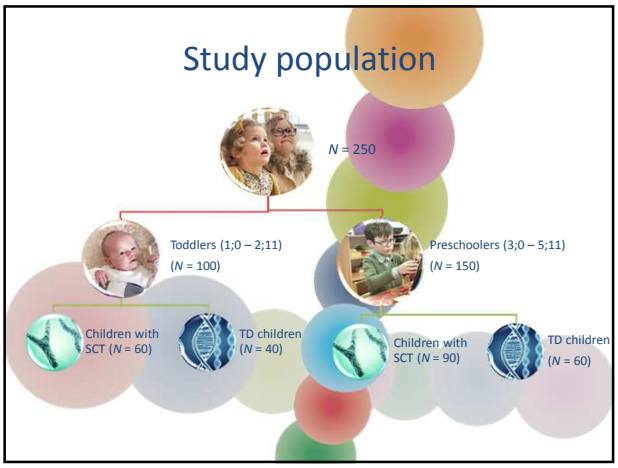
2016:
personal grant from the Dutch government (800.000 Euros = +/- $ 900.000), awarded to Sophie van Rijn

- ✓ Cognitive and behavioral development of children with SCT aged 1 to 6 years
- ✓ Improve clinical care: prevention and intervention

- ➢ **Focus on risk emotional, social and behavioral problems**
- ➢ **Understand the underlying mechanisms in terms of how the brain processes information**

The study started in November 2016: we are up and running!

Study population



Why focus on social, emotional and behavioral problems?

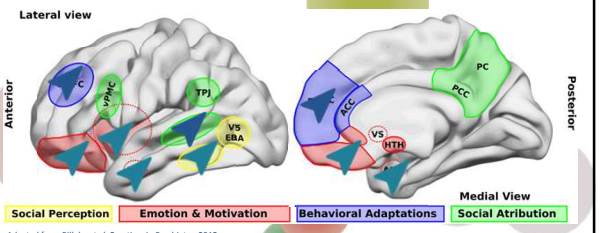
	Children with an extra X (34 boys and 26 girls 8-18 yrs)		
CBCL	Average (T<65)	Mild (65<T<70)	Severe (T>70)
Social problems	58.5 %	24.5 %	17.0 %
Attention problems	71.7 %	9.4 %	18.9 %
Thought problems	62.3 %	22.6 %	15.1 %
Anxious - depressed	71.7 %	15.1 %	13.2 %
Withdrawal	62.3 %	22.6 %	15.1 %
Somatic complaints	73.6 %	9.4 %	17.0 %
Aggressive behavior	84.9 %	11.3 %	3.8 %
Rule breaking behavior	88.7 %	9.4 %	1.9 %

(Van Rijn et al., 2015)

Parts of the brain important for social-emotional processing

15 adults with XXY + controls
18 children with XXY (8-18 yrs) + controls

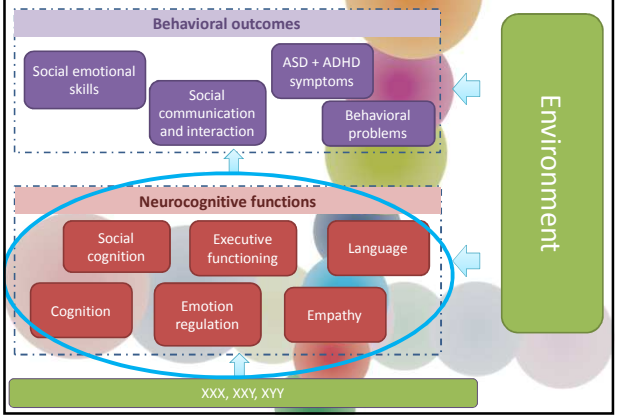
Functional MRI, VBM, DTI, Resting state MRI

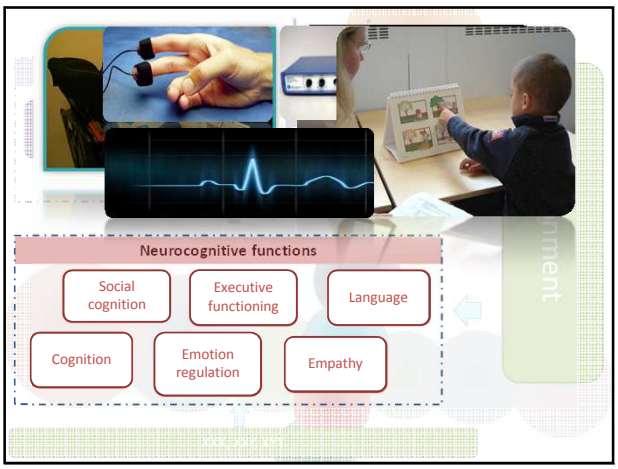


Adapted from Billeke et al. Frontiers in Psychiatry, 2015

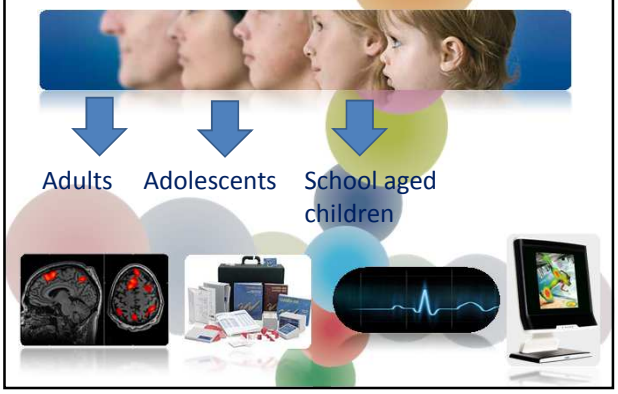
(Raznahan et al., 2014; Brandenburg-Goddard et al., 2014, 2015, 2016 ; Van Rijn et al. 2008, 2012)

Research domains





Our research so far:



Adults, adolescents and school aged children show vulnerabilities in:

Communicative skills

Express your thoughts
Understand what others are saying
Rely on gestures and tone of voice
Read 'between the lines'

(Van Rijn et al., 2006; Van Rijn et al., 2011; Bierman et al., submitted)

Adults, adolescents and school aged children show vulnerabilities in:

Executive skills (cognitive control)

Focus and concentrate of what's important
Filter out irrelevant information
Re-adjust and reconsider: flexibility
Inhibit inappropriate responses
Manage complex and conflicting information

(Van Rijn et al., 2015)

Adults, adolescents and school aged children show vulnerabilities in:

Social cognitive skills

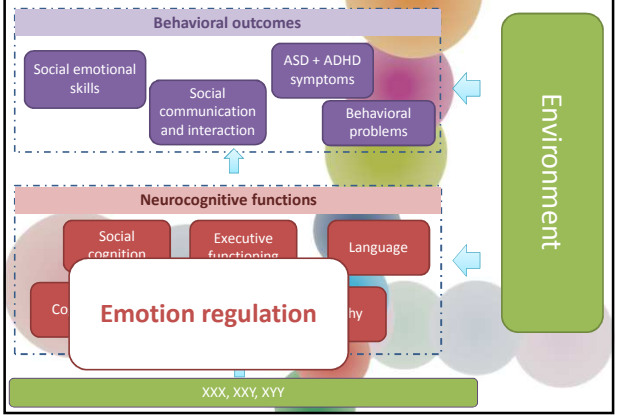
Understand facial expressions
Read body language
Understand perspectives of others
Social orienting
Social attention

(Van Rijn et al., 2006; Van Rijn et al., 2014; Van Rijn et al., 2015)

Our current research:

Adults | Adolescents | School aged children | Young children

Research domains



Emotion regulation

Emotion
=
communication

Emotion regulation

Definition by Gross, 1998

"process by which we influence
what emotions
we experience
when
and *how*
we express them"

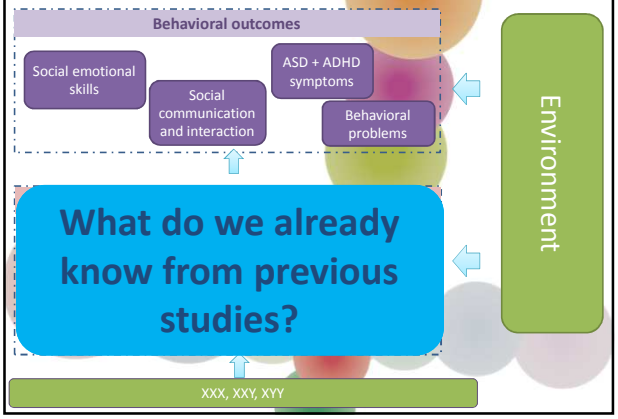
Development of emotion regulation

- Shift from automatic to effortful
- Social interactions needed
- Learn by observation and doing
- Different skills necessary
  - Language
  - Executive functions
  - Social cognition
- Bodily reaction

What does it look like?

- On the outside:
  - Easily frustrated / out of balance
  - From 0 to 100 in seconds
  - Tend to react directly (hitting, biting, screaming)
  - Tantrums / fights with others
- On the inside:
  - Easily upset
  - Worry / anxious / sadness
  - Withdrawal from social situations
  - Turning away from parent(s)

Research domains



Emotion awareness

- Alexithymia: inability or reduction to identify, experience, describe and reflect on one's own emotions (Lane et al., 1997) → emotion regulation
- BVAQ: alexithymia questionnaire

(Van Rijn et al. 2006)

Results

N = 32 men with XXY

Dimension of alexithymia	Controls	Men with Klinefelter
Verbalizing emotions	20.2 (6.2)	24.6 (8.3)*
Fantasizing	24.6 (8.2)	21.1 (8.0)
Identifying emotions	15.6 (4.0)	20.2 (7.0)*
Emotionalizing	26.2 (6.3)	20.7 (5.9)*
Analyzing emotions	18.6 (6.1)	19.2 (7.7)

More problems with verbalizing emotions

More problems with identifying emotions

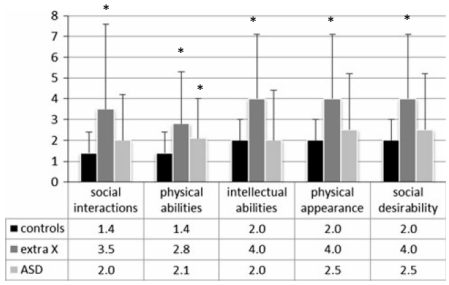
More easily aroused

Social anxiety

SAS: Dutch questionnaire to assess cognitive and affective reactions in social situations

Participants: 34 XXY, 26 XXX, 58 ASD, 106 controls.

(Van Rijn et al. 2014)



Children with an extra X have more social anxiety than children with ASD

Increased emotion regulation problems?

Emotion regulation

- Cognitive emotion regulation questionnaire (CERQ)
- Self-report

9 subscales:
- *Self blame*
- *Other-blame*
- *Rumination / focus on thoughts*
- *Catastrophizing*
- *Putting into perspective*
- *Positive refocusing*
- *Positive reappraisal*
- *Acceptance*
- *Refocus on planning*

Cognitive coping strategies to deal with threatening or stressful events

(Bierman et al. submitted)

- *N* = 144: 23 girls with XXX, 30 boys with XXY, and 91 controls
- Age: 12.6 (*SD* = 3.1)

Subscale CERQ	Controls	Children with extra X
Rumination	9.6 (3.9)	11.0 (3.9)*
Blaming others	6.5 (2.6)	7.8 (3.0)*

Predictors for problem behavior (CBCL):

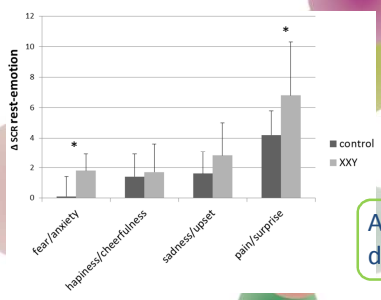
- Somatic concern
- Emotional outbursts

- Somatic concern
- Rule breaking behavior
- Withdrawal

Other subscales: no differences between children with extra X and controls

Physiological arousal: skin conductance

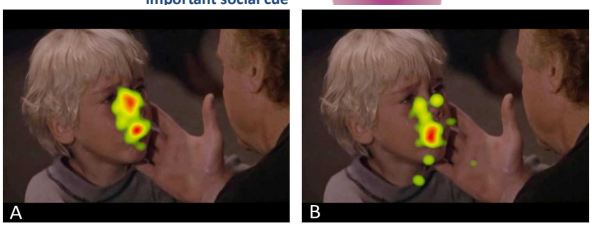
- In adults: 17 adults with XXY



Attentional deployment?

Eye tracking: social attention

Eyes: most important social cue



'Emotion-dysregulation-hypothesis'

social interaction
↓
difficulties in understanding social signals
difficulties in regulating emotions (executive dysfunction)
↓
increased arousal
↓
avoidance of eyes
↓
insufficient down-regulation of arousal
↓
anxiety and distress during social interactions
↓
reduced social learning experiences

(Van Rijn et al., 2014)

Self-management training

- Self management = the ability to regulate emotions and behavior, adaptive to the environment

Effects on an individual's ability to tolerate unmet wants or needs, handle disappointments and failures, and work towards success
(Bandy & Moore, 2010)

Training

- Psycho-education
- Personal goals
- Improving insight in personal neurocognitive profile
- Skills training for dealing with social and emotional problems
- Relaxation exercises
- Transfer to daily life: weekly assignments, workbook

Sessions: 5 months, 10 sessions, 90 minutes

Effectivity of self-management training

16 adults with XXY

Methods: questionnaires pre-test and post-test
Self report and informant report

Emotional / behavioral
Autism like behaviors
Social skills

→ **Self report**
Informant report

(Martin et al., submitted)

Results

- Significant increased awareness of limitations in social flexibility and communication

  Important for: learning alternative strategies, adapting behavior, improve coping

- Significant less fear, depression and emotional outburst

  Important for: becoming more assertive, positive interactions, to solve interpersonal conflicts more constructively

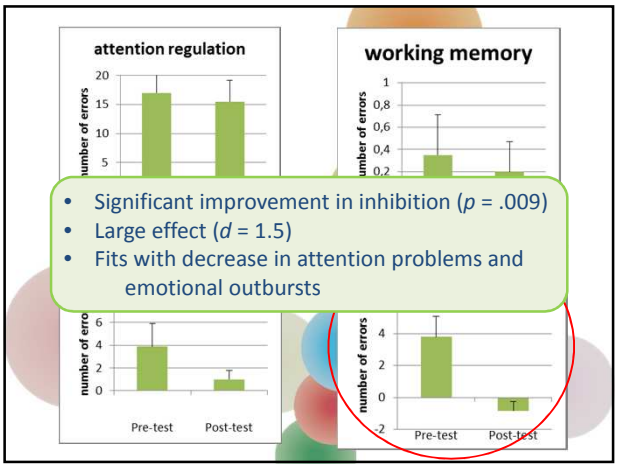
- Significant less attention problems (less distracted and impulsive) → more control

Effectivity of self-management training (2)

Cognitive assessment (*N* = 14)

- pre-test and post-test

ANT test battery
Inhibition
Working memory
Mental flexibility
Attention regulation

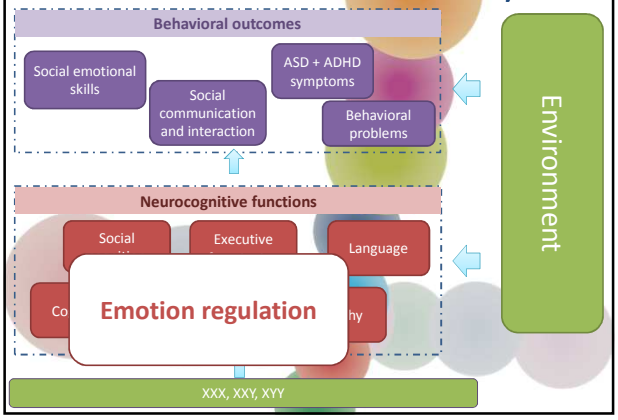


Emotion regulation: What's next?

- Vulnerable domain in adults SCT
- Strong associations with psychological problems in general population (Aldao et al., 2010)

- How about (young) children with SCT?
- Relation with other cognitive functions?
- Risk for behavioral difficulties?

Research domains of TRIXY Study



International collaboration

Thank you!