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

Leiden University – The Netherlands

TRIXY Center of Expertise
Trisomy of the X and Y Chromosomes

Leiden University – Treatment and Expertise Center
Leiden University – Medical Center

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

- Toddlers (1;0 – 2;11) (N = 100)
- Preschoolers (3;0 – 5;11) (N = 150)
- Children with SCT (N = 60)
- TD children (N = 60)

Why focus on social, emotional and behavioral problems?

- Why focus on social, emotional and behavioral problems?

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

- Social Perception
- Emotion & Motivation
- Behavioral Adaptation
- Social Attribution

Research domains

- Social emotional skills
- Language
- Social cognition
- Executive functioning
- Emotion regulation
- Empathy

Neurocognitive functions

- Social cognition
- Executive functioning
- Language
- Emotion regulation
- Empathy

Our research so far:

- Adults
- Adolescents
- School aged children
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)

**Executive skills (cognitive control)**
- Focus and concentrate on what's important
- Filter out irrelevant information
- Re-adjust and re-assess: flexibility
- Inhibit inappropriate responses
- Manage complex and conflicting information

(Van Rijn et al., 2015)

**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

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**Research domains**

Behavioral outcomes
- Social emotional skills
- Social communication and interaction
- ASD + ADHD symptoms
- Behavioral problems

Neurocognitive functions
- Social cognition
- Executive functions
- Language

Emotion regulation
- Emotion = communication

XXX, XXY, XYY

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Environment
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)

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

Research domains

Behavioral outcomes

Social emotional skills

Social communication and interaction

ASD + ADHD symptoms

Behavioral problems

What do we already know from previous studies?

Cognition

Social

communication and interaction

ASD + ADHD

symptoms

Behavioral problems

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

Results

N = 32 men with XXY

<table>
<thead>
<tr>
<th>Dimension of alexithymia</th>
<th>Controls</th>
<th>Men with Klinefelter</th>
</tr>
</thead>
<tbody>
<tr>
<td>Verbalizing emotions</td>
<td>20.2 (6.2)</td>
<td>24.6 (8.5)*</td>
</tr>
<tr>
<td>Fantasizing</td>
<td>24.6 (8.2)</td>
<td>21.4 (8.0)</td>
</tr>
<tr>
<td>Identifying emotions</td>
<td>15.6 (4.0)</td>
<td>20.2 (7.0)*</td>
</tr>
<tr>
<td>Emotionalizing</td>
<td>20.2 (8.3)</td>
<td>20.7 (5.9)*</td>
</tr>
<tr>
<td>Analyzing emotions</td>
<td>18.6 (8.1)</td>
<td>20.2 (8.0)</td>
</tr>
</tbody>
</table>

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)

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

(Bierman et al. submitted)

Physiological arousal: skin conductance
- In adults: 17 adults with XXY

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

(Bundy & 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

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

- ANI test battery
  Inhibition
  Working memory
  Mental flexibility
  Attention regulation
Significant improvement in inhibition ($p = .009$)
Large effect ($d = 1.5$)
Fits with decrease in attention problems and emotional outbursts

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

- Behavioral outcomes
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Environment

Emotion regulation

International collaboration

Thank you!