Narrative comprehension and production abilities of children with 22q11.2 deletion syndrome

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ABSTRACT

Background: The 22q11.2 Deletion Syndrome (22q11DS) is associated with language deficits and weak intellectual functioning. In other clinical groups, linguistic and cognitive difficulties have been associated with impaired acquisition of narrative abilities. However, little is known about the narrative abilities of children with 22q11DS.
Aims: To describe the ability of children with 22q11DS to produce and comprehend narrative macrostructure. Additionally, to examine the role of intellectual functioning in explaining their narrative difficulties.
Methods and procedures: Narrative skills of 14 school-aged children with 22q11DS were compared to those of younger typically developing (TD) children matched on mental age and same-aged peers with Developmental Language Disorder (DLD).
Outcomes and results: Children with 22q11DS had significantly lower scores on narrative comprehension than younger TD children. No significant differences emerged on narrative production. Children with 22q11DS and children with DLD did not differ significantly on any of the narrative measures.
Conclusions and implications: Narrative comprehension in children with 22q11DS seems more affected than production. Narrative comprehension difficulties cannot be entirely explained by a low level of intellectual functioning. Narrative comprehension and production abilities in 22q11DS require further consideration.

What this paper adds

Children with 22q11.2 Deletion Syncrome (22q11DS) are reported to present with impairments across all domains of language,

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including impaired narrative abilities. However, evidence regarding the kind of narrative difficulties that are associated with 22q11DS is limited, and to date studies have only focused on difficulties in narrative production. This study therefore examines both productive and receptive narrative abilities in children with 22q11DS. Moreover, as most children with 22q11DS function on a borderline intellectual level, this study compares narrative abilities of children with 22q11DS to those of typically developing children matched for mental age. This comparison helps to answer the question whether narrative difficulties of children with 22q11DS exceed their developmental delay and therefore warrant more attention in research and clinical care. In addition, we know from children with Developmental Language Disorder (DLD) that narrative difficulties can occur in absence of intellectual problems. Therefore, we also compared narrative abilities of children with 22q11DS to those of children with DLD. By comparing these groups with one another we can enhance our understanding of which (linguistic) mechanisms could play a role in narrative difficulties in 22q11DS.

1. Introduction

The acquisition of narrative abilities, the abilities to use language to tell and understand stories, is difficult for children who experience language problems (Fey, Catts, Proctor-Williams, Tomblin, & Zhang, 2004). Difficulties in narrative development may negatively affect a child's daily functioning, as narrative abilities are essential to communicate with peers and family, and to interact in a school context (Boudreau, 2008). The 22q11.2 deletion syndrome (22q11DS) is a genetic disorder that is, amongst others, associated with severe difficulties in language development (for an overview, see Solot et al., 2019). Many children with 22q11DS also experience problems in social interaction as well as learning difficulties (Swillen & McDonald-McGinn, 2015). Weak narrative abilities may play a role in the occurrence of such problems in 22q11DS. Therefore, the first aim of the present study is to investigate the narrative production and comprehension abilities of children with 22q11DS.

We compare the narrative abilities of children with 22q11DS with those of typically developing (TD) children matched on mental age. Hereby, we investigate whether narrative abilities of children with 22q11DS lag behind the level of what can be expected for their level of cognitive development, as a below average level of intellectual functioning is characteristic for 22q11DS. In addition, we compare the narrative abilities of children with 22q11DS to those of chronologically age-matched children with a Developmental Language Disorder (DLD). This comparison allows us to study the role of intellectual functioning in the narrative abilities of children with 22q11DS, as, in contrast to 22q11DS, DLD is mostly associated with an average level of intellectual functioning (Bishop, Snowling, Thompson, Greenhalg, & the CATALISE-2 consortium, 2017; Swillen & McDonald-McGinn, 2015).

1.1. The 22q11.2 deletion syndrome

22q11DS is caused by a micro-deletion on the long arm of chromosome 22 and is estimated to occur in 1 in 3,000–6,000 (live) births (McDonald-McGinn et al., 2015). The syndrome has many possible symptoms, which can vary in their expression across individuals and affect almost any part of the body. Common symptoms include congenital heart defect, palatal abnormalities and intellectual impairment. Most children with 22q11DS function on a level of borderline intelligence (IQ between 70–85) or mild intellectual disability (IQ between 55–70; De Smedt et al., 2007; Swillen, Moss, & Duijff, 2018). Additionally, children with 22q11DS are at increased risk for psychopathology, especially Autism Spectrum Disorder, Attention Deficit Hyperactivity Disorder, and Anxiety Disorders in childhood, and schizophrenia in young adulthood (Fiksinski et al., 2018).

One of the earliest developmental symptoms noted by parents of children with 22q11DS and clinicians is the delayed achievement of language milestones (Solot et al., 2019). Over 90 % of children with 22q11DS do not become verbal within the typical age limits (Gerdes et al., 1999; Mills, Gosling, & Sell, 2006; Solot et al., 2000). Over the course of childhood, the majority of children with 22q11DS continue to have difficulties across various language domains, such as vocabulary and grammar (Glaser et al., 2002; Persson, Niklasson, Oskarsdottir, Johansson, Jönsson, & Söderpalm, 2006; Solot et al., 2019; Van den Heuvel, Manders, Swillen, & Zink, 2018). In addition, while typically developing (TD) children generally have better receptive language skills (understanding language) compared to expressive language skills (producing language; Bates et al., 1993), this advantage of receptive language skills seems smaller in school-aged children with 22q11DS (Van den Heuvel et al., 2018). This suggests that monitoring receptive language skills in these children may thus be particularly important.

Finally, children with 22q11DS often struggle to effectively use language in a social context, as is evident from problems in communication, interaction and peer relations in children with 22q11DS (Angkustsiri et al., 2014; Persson et al., 2006; Van den Heuvel, Manders, Swillen, & Zink, 2017). One aspect of language that is especially important for language use in everyday life is the development of narrative abilities (Botting, 2002), which is the focus of the current study.

1.2. Development and assessment of narrative abilities

A narrative is a story that is used to inform others about a sequence of personal or fictional events in a coherent and structured way. Both our ability to understand and to produce narratives is vital, given their prominent role in social interactions in personal, school and formal settings. Moreover, the development of narrative abilities is related to the development of literacy skills and academic success (Botting, 2002; Johnston, 2008; Westerveld & Gillon, 2010). Studies in the general population show that children's narrative development starts around the age of 2 years and continues into adolescence. Around the age of 9 years old, most children are able to tell a story that connects a series of actions and events, contains a coherent plot, and involves character descriptions (Pinto, Tarchi, & Bigozzi, 2019; Roelofs-Borgers, 1998).

To assess story generation abilities, a child is usually requested to tell a story using a set of pictures as prompts. The produced

narrative can be analyzed globally at the level of narrative macrostructure by evaluating organizational aspects of the narrative that support transfer of story content, such as the use of an episodic structure. In addition, narrative production can be analyzed locally at the level of narrative microstructure by evaluating the linguistic aspects of the narrative, such as lexical diversity and grammatical complexity. To assess narrative comprehension, a child is requested to answer a set of comprehension questions about the story events and emotional states of the characters (Botting, 2002; Norbury & Bishop, 2003).

1.3. Narrative problems and associated mechanisms

For some children, the development of narrative production and comprehension lags behind in comparison to their peers. A well-known group that experiences persistent narrative difficulties are children with a diagnosis of Developmental Language Disorder (DLD). Around 5–7 % of children in the general population receive this diagnosis, because they have severe problems in language acquisition in absence of intellectual disability and without an evident physical, neurological or environmental cause (Bishop et al., 2017). Research on the narrative abilities of children with DLD has shown that their difficulties in narrative production and comprehension are associated with impairments in (a combination of) linguistic, cognitive and/or social functions (Blom & Boerma, 2016; Duinmeijer, de Jong, & Scheper, 2012; Lindgren, 2019; Lynch et al., 2008; Matthews, Biney, & Abbot-Smith, 2018), depending on which specific narrative skills are evaluated (Duinmeijer et al., 2012). Difficulties in the organization of plot structure and transfer of story content (i.e., in production of narrative macrostructure) are reported to be relatively independent of language ability, including receptive vocabulary size and grammar knowledge (Blom & Boerma, 2016; for a discussion, see Boerma, Leseman, Timmermeister, Wijnen, & Blom, 2016). Rather, weak production of narrative macrostructure is associated with impairments in cognitive functions, such as attention, working memory and the use of real-world knowledge to understand a (social) situation (Blom & Boerma, 2016; Duinmeijer et al., 2012; Ketelaars, Jansonius-Schultheiss, Cuperus, & Verhoeven, 2012). This contrasts with weak performance on measures of narrative microstructure, which is often associated with lower language skills in children with DLD (Botting, 2002). Finally, problems with narrative comprehension in these children have been related to poorer receptive vocabulary, weaker sustained attention and inference problems (Blom & Boerma, 2016; Boerma et al., 2016).

1.4. Narrative skills of children with 22q11DS

Children with 22q11DS may be specifically vulnerable to develop difficulties in production of narrative macrostructure and narrative comprehension, given that 22q11DS is associated with language problems as well as impairments in the cognitive and social skills that have been associated with narrative difficulties in DLD. Solot et al. (2019) summarized the following narrative difficulties in children with 22q11DS: "*Extracting salient points from verbal or written narrative, understanding implications, making inferences and predictions, and use of disorganized, terse, ambiguous, or verbose narratives*" (p.988). To our knowledge, only two studies have directly assessed the narrative skills of children with 22q11DS (Persson et al., 2006; Van den Heuvel, Reuterskjöld, Solot, Manders, Swillen, & Zink, 2017). Persson et al. (2006) used the Bus Story retelling task with 19 school-aged children with 22q11DS. The children did not make more grammatical errors compared to TD children of the same age, but produced shorter and fewer grammatically complex sentences. In addition, children with 22q11DS needed more encouragement to take initiative and they transferred less essential information in their stories. Van den Heuvel, Reuterskjöld et al. (2017) assessed 27 children with 22q11DS between 6 and 14 years old with a perspective taking task, in which children were asked to describe a picture and ascribe feelings and thoughts to the characters. They report that children with 22q11DS provided much information about irrelevant visual details, resulting in a chain of unconnected utterances. The authors conclude that children with 22q11DS transferred less essential information than TD children. Taken together, emerging evidence suggests that children with 22q11DS indeed experience problems in the production of narratives. However, more research is warranted, in particular with regard to the comprehension aspect of narrative skills and whether narrative abilities of children with 22q11DS are in line with what can be expected for their level of cognitive development.

1.5. Research aims and hypotheses

Given that the development of narrative abilities is critical for communicating personal experiences, social interaction and academic functioning, a better understanding of the narrative production and comprehension abilities in 22q11DS is important. Narrative skills have been shown to build on linguistic and cognitive functions (Johnston, 2008; Matthews et al., 2018); this holds especially for production of narrative macrostructure and narrative comprehension (Blom & Boerma, 2016). Consequently, children with 22q11DS are at a high risk of impairment in these domains, as this syndrome is associated with an increased risk for both intellectual and linguistic difficulties. Therefore, the first aim of the present study is to describe the narrative abilities of children with 22q11DS. Subsequently, we compare the narrative abilities of children with 22q11DS with those of a group of younger TD children matched on mental age. This comparison will show if their narrative abilities are keeping with, or impaired beyond, what may be expected for their developmental level. We expect children with 22q11DS to perform on par with their younger, mentally age-matched peers on the measures for production of narrative macrostructure and narrative comprehension. If confirmed, this would indicate that their delay in narrative development is in keeping with their global cognitive development.

Furthermore, we compare the narrative abilities of children with 22q11DS with a group of same-aged peers with a diagnosis of DLD. We may expect that children with 22q11DS perform on par with the children with DLD, despite the overall higher level of intellectual ability in the DLD group, as we know that children with DLD can demonstrate weak language skills in the absence of intellectual problems. If confirmed, this may indicate that narrative difficulties of children with 22q11DS cannot entirely be attributed to

a lower level of intellectual functioning However, it could also be that such a discrepancy between intellectual ability and language skills is a unique feature of DLD. Alternatively, the children with DLD could perform better than the same-aged children with 22q11DS on the narrative tasks, given that the latter group has both intellectual and linguistic difficulties. This could suggest a role of specific language difficulties in addition to low intellectual ability in narrative abilities in 22q11DS.

2. Method

2.1. Participants

Our participants took part in a larger project which aimed to measure brain activation during language processing by using brain scans (fMRI; Vansteensel et al., 2021). A total of 14 children with 22q11DS and 15 children with DLD, all between 6–10 years old, were included in this study. For both groups, we only included children who did not present with intellectual disability (verbal or non-verbal IQs were higher than 70), hearing loss (>35 dB) and a diagnosis of Autism Spectrum Disorder. Parents of all participants gave written informed consent for their child to participate in the study. The study was approved by the Ethical Review Board of the University Medical Center Utrecht, and was performed in accordance with the Declaration of Helsinki (2013).

Narrative task results of children with 22q11DS and children with DLD were compared with data from 14 younger TD children, who were selected from a larger pool of children that participated in an earlier study (Boerma et al., 2016). We matched the TD children to our participants with 22q11DS based on their nonverbal mental age, since we aimed to investigate whether narrative abilities of 22q11DS children were in line with their developmental level. For all TD children, intelligence scores of the short version of the Wechsler Nonverbal-NL (WNV; Wechsler & Naglierei, 2008) were available. For both children with 22q11DS and children with DLD, we collected intelligence scores from standardized intelligence measures (mostly Wechsler tests) obtained from either medical or school records. If intelligence was assessed more than two years prior to the study, we administered the short version of the WNV-NL at the start of the test session. We calculated the mental age of all participants using the formula: *[(full scale IQ score / participants chronological age) * 100]* (Caplan, Neece, & Baker, 2015). Participant characteristics are presented in the results section (see Table 1).

2.2. Measures and scoring

2.2.1. Language abilities

We used two measures to collect background information on the language abilities of our participants with 22q11DS and DLD: 1) To assess children's grammatical language skills, we used the sentence repetition subtest of the Dutch adaptation of the Clinical Evaluation of Language Fundamentals (CELF-IV-NL; Kort, Compaan, Schittekatte, & Dekker, 2010). For this task, children are asked to repeat sentences of increasing difficulty, read to them by a researcher. This task is often used to identify children with DLD (Klem et al., 2015). We used children's chronological age to convert raw scores into age-corrected standard scores (M = 10; SD = 3) with a higher raw score indicating better grammatical skills and a standard score below 7 indicating "below average performance"; 2) to assess receptive vocabulary, we used the Dutch adaptation of the Peabody Picture Vocabulary Test (PPVT-III-NL; Schlichting, 2005). During this task, children are shown four different pictures and are requested to point to the picture that corresponds to the target word that is read out loud by the researcher. Performance on the PPVT is measured as a quantitative score with higher raw scores indicating better word comprehension skills. We converted raw scores into age-corrected standard scores (M = 100; SD = 15). PPVT scores were also available for the group of TD children.

2.2.2. Narrative abilities

Children's abilities to produce and comprehend narrative macrostructure were measured with the Multilingual Assessment Instrument for Narratives (MAIN; Gagarina et al., 2012). The MAIN was developed within the framework of the COST Action IS0804 *Language Impairment in a Multilingual Society: Linguistic Patterns and the Road to Assessment* and can be used to assess different aspects of narrative comprehension and production of (bilingual) children from 3 to 10 years old. For the purpose of the current study, we used a *model story* and a *production story* from the MAIN stimulus set. Both stories are depicted by a sequence of six pictures and contain three story episodes. In each episode, elements of the time and place of the story and story characters are introduced. Furthermore, each story episode contains a goal (e.g., cat wanted to catch baby birds), an attempt to reach that goal (e.g., cat tried to climb the tree) and an outcome of that attempt (e.g., cat was chased away by the dog). In addition, each story episode includes elements that are related to the internal/mental state of the main characters (e.g., cat was scared).

All children first saw and heard the *model story* (Cat), which was read to them by the researcher, and were asked ten comprehension questions that targeted the story structure and internal states of the characters. Hence, the main goal of using the model story was to introduce the narrative assessment, and to evaluate narrative comprehension. Subsequently, all children were asked to generate their own story using the stimulus set that belongs to the *production story* (Baby birds) to assess production of narrative macrostructure. This was followed by a similar set of ten comprehension questions. Most children enjoyed this narrative task, which was administered as the final task of a test session of about 60 min in which children completed several language and cognitive tasks.

2.2.3. Scoring narrative abilities

We used the standard outcome measures for production and comprehension of narrative macrostructure offered by the MAIN: 1) Production. We counted how many story structure elements children incorporated during telling of the production story. The inclusion of these story elements is awarded points, resulting in a production score with a higher score indicating a better performance. The story

Table 1

Means and standard deviations for age (in months), IQ scores and standardized language measures for the children with DLD, children with 22q11DS and TD children.

Variable	TD			22q11DS			DLD			Comparison	
	N	M	SD	N	M	SD	N	M	SD	χ^2/F	p
Gender (males)	7			8			8			0.14	.931
Chronological age	14	77.2	14.7	14	104.2	19.1	15	98.4	20.6	8.40	.001
Mental age	14	76.6	14.2	14	76.7	14.6	14	104.7	23.9	11.18	<.001
Total IQ score	14	99.4	6.0	14	74.0	8.6	14	105.4	15.8	32.37	<.001
PPVT raw score	14	92.2	13.0	13	88.5	8.4	15	95.1	14.4	1.00	.376
PPVT standard score	14	108.2	8.2	13	83.1	13.7	15	93.2	13.6	14.86	<.001
CELF raw score	–	–	–	14	33.2	10.1	15	25.1	12.1	3.80	.062
CELF standard score	–	–	–	14	5.2	2.2	15	3.9	2.0	−1.64	.113

Note. One girl with 22q11DS did not complete the PPVT. For one girl with DLD we were not able to compute her mental age, because we did not manage to obtain the IQ information from her school. *PPVT: Peabody Picture Vocabulary Test; *CELF: the sentence repetition task of the Clinical Evaluation of Language Fundamentals.

elements included the setting (0, 1 or 2 points) and, for each episode, the internal state as initiating event, goal, attempt, outcome, and the internal state as reaction (each 0 or 1 point; max. 17 points). 2) Comprehension of the model story and production story. For each story, the MAIN provides ten comprehension questions to assess children's understanding of the story events and their consequences, and the goals of the main characters and their thoughts and feelings. For each story separately, children were awarded one point for each question that they answered correctly. Hence, a higher score indicates a better story comprehension (each story max. 10 points). A high quality microphone (Samson Go Mic) was used to record all narratives. The narratives were scored offline by a trained researcher. Over 40 % of the narratives from the TD group were scored by a second independent rater, resulting in acceptable inter-rater agreement (for exact numbers see Boerma et al., 2016). The data of children in the two clinical groups were all scored using the same protocol. In case of uncertainties, a final score was awarded by consensus.

2.3. Data analyses

All analyses were performed in SPSS version 26. We confirmed our matching of the children with 22q11DS to TD children, based on mental age by using Univariate Analyses of Variance (ANOVA). In addition, we compared the three groups on chronological age, intelligence and gender distribution by using ANOVA and χ^2 tests. Given that significant differences in chronological age and intelligence are inherent to the design of this study, we only adjust for possible gender differences in our analyses. In addition, we evaluated the language abilities of children with 22q11DS and children with DLD using their raw and standard scores on the PPVT and the sentence repetition task.

The main aim of the current study was to compare narrative production and comprehension between children with 22q11DS, TD children and children with DLD. Since the MAIN narrative assessment does not provide age-corrected standard scores, we took the performance of the group of TD children as a reference to evaluate the narrative performance of children with 22q11DS and children with DLD. Given our limited sample size and the fact that we measured narrative abilities on an ordinal scale, we used three non-parametric Kruskal-Wallis tests with respectively the narrative production score, the comprehension score on the model story and the comprehension score on the production story as the outcome variables and with group (22q11DS, DLD, TD) as the independent variable. The Kruskal-Wallis test is a rank-based test, implying that scores of all children are ordered from lowest to highest and are assigned a rank. We therefore considered it insightful to display the median score for each group in our sample descriptives. If the Kruskal-Wallis test resulted in a significant group difference on any of the narrative measures, we used non-parametric Mann-Whitney tests for pairwise comparisons and applied Bonferroni correction for multiple testing. We additionally calculated the effect size r, by dividing the z-score of each pairwise comparison (as provided by SPSS) by the square root of the total number of observations. We visually inspected the frequency distribution of number of points that children received on these narrative measures.

Our limited sample size and choice of narrative measure prevent us from drawing meaningful conclusions regarding the associations between our measures for linguistic and intellectual functioning and narrative abilities. We therefore included the results of these analyses as supplementary material (see Appendix A).

3. Results

3.1. Matching procedure and participant characteristics

Table 1 shows the demographic information of the participants as well as their scores on intelligence and standardized language tests for each group and the statistics of these variables. We successfully matched the children with 22q11DS to the TD children based on their mental age (p = 1.00). IQ scores of the children with 22q11DS fell on average in the borderline range, significantly lower than those of the TD children and children with DLD (p < .001), implicating that chronological and mental age differed significantly between the three groups. That is, the children with 22q11DS had a significantly higher chronological age than the TD children (p = .001), but did not differ from the children with DLD in chronological age (p = 1.00). Moreover, the children with 22q11DS and TD

Table 2
The means, standard deviations, medians, and ranges for the score on the production task for the children with 22q11DS, children with DLD, and TD children.

Group	N	Mean	SD	Median	range
TD	14	8.6	2.2	8.5	5 - 12
22q11DS	14	7.0	2.6	7.0	2 - 12
DLD	15	7.2	2.5	6.0	4 - 13

Table 3
The means, standard deviations, medians and ranges for the comprehension score for the model story and the production story for the children with 22q11DS, children with DLD, and TD children.

	Model story					Production story				
Group	N	Mean	SD	Median	Range	N	Mean	SD	Median	Range
TD	14	9.6	0.5	10.0	9−10	14	9.0	1.2	9.5	7−10
22q11DS	14	8.4	1.4	9.0	6−10	13	6.5	1.9	6.0	4−10
DLD	15	8.6	1.4	9.0	5−10	14	7.4	1.9	8.0	5−10

Note. Comprehension data for the production story were not available for 1 child with 22q11DS and 1 child with DLD due to technical issues during data collection.
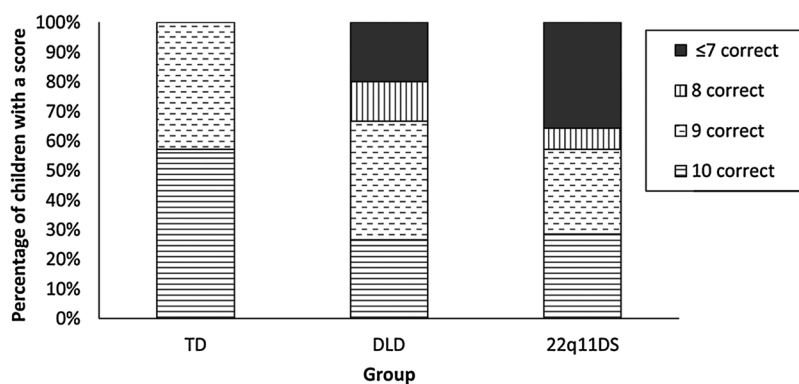

Fig. 1. The proportion of children per group with respectively 10 (= max), 9, 8, or less than 8 answers correct on the comprehension questions of the model story.

children had a significantly lower mental age as compared to the children with DLD ($p = .001$). Finally, the children with DLD did not differ from children with 22q11DS with regard to their scores on the background language measures and both groups did not differ from the TD children on the raw score of the PPVT. Children with 22q11DS scored more than 1 standard deviation (SD) below the mean on the PPVT, whereas children with DLD performed within the average range on this task. For the sentence repetition task, both children with 22q11DS and children with DLD obtained on average a standard score lower than 7, indicating a below average performance.

3.2. Narrative performance

3.2.1. Narrative production

Table 2 shows the means, standard deviations, medians and range of participant scores per group on the narrative production task. The scores did not differ significantly between the children with 22q11DS, children with DLD and children in the TD group, indicating that we did not detect evidence for a difference between children in the number of story structure elements they produced [$H(2) = 3.74, p = .154, \eta^2 = .01$; see Table 2].

3.2.2. Narrative comprehension

Significant differences between the three groups were observed for comprehension of both the model and the production story (see Table 3).

Model story. The Kruskal-Wallis test showed a significant main effect of group for the comprehension score of the model story [$H(2) = 6.58, p = .037, \eta^2 = .06$]. Additional pairwise comparisons did not survive Bonferroni correction. However, we found a medium effect size both for the comparison between children with 22q11DS and TD children ($U = 52.00, p = .024, r = 0.43$) and between children with DLD and TD children ($U = 58.00; p = .027, r = 0.41$). Finally, the observed small difference between 22q11DS and DLD
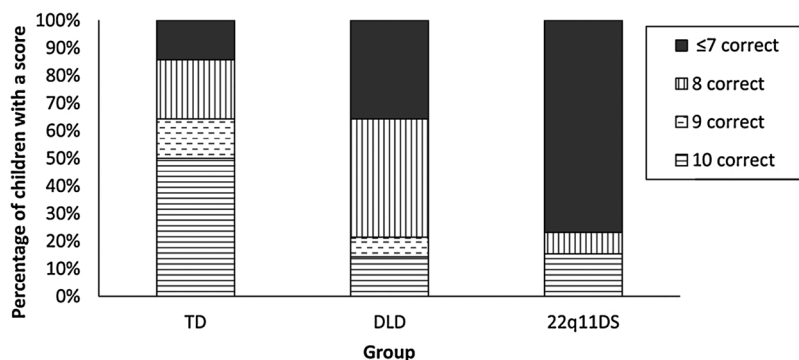
Fig. 2. The proportion of children with respectively 10 (= max), 9, 8, or less than 8 answers correct on the comprehension questions of the production story.

was not statistically significant (U = 97.00; $p =$ .717, r = 0.07). Fig. 1 displays the distribution of scores per group on the comprehension task for the model story. Visual inspection of the data shows that the TD children performed at or near ceiling level, as eight children (57 %) answered all questions correctly and six children (43 %) only gave one incorrect answer. For the children with 22q11DS, only four children answered all questions correctly (29 %) and six children provided more than one incorrect answer (43 %). Similarly, only four children with DLD answered all questions correctly (27 %) and five children provided more than one incorrect answer (33 %; see Fig. 1).

Production story. The Kruskal-Wallis test showed a significant main effect of group for the comprehension score of the production story [H(2) = 12.02, $p =$ .002, $\eta^2 = 0.21$]. Pairwise comparisons showed that scores of the children with 22q11DS differed significantly from those of the TD children, with a large effect size (U = 26.50, $p =$ .001, r = 0.62), indicating that the children with 22q11DS gave fewer correct answers to the comprehension questions of the production story. Again, the comparison between the comprehension scores of the children with DLD and the TD children did not survive Bonferroni correction, although we found a medium effect size (U = 49.00, $p =$ .020, r = 0.44). Finally, the comprehension score did not differ between the children with 22q11DS and children with DLD (U = 59.00, $p =$ .114, r = 0.22). Fig. 2 displays the distribution of scores per group on the comprehension task for the production story. Seven children of the TD group answered all comprehension questions correctly (50 %), and the two weakest TD children provided three incorrect answers (14 %). Only two children with 22q11DS (15 %) answered all questions correctly and eight children provided more than three incorrect answers (62 %). Only two children with DLD answered all questions correctly (14 %) and four children provided more than three incorrect answers (29 %; see Fig. 2).

4. Discussion

In this study, we investigated the ability of school-aged children with 22q11DS to produce and comprehend narratives. We evaluated children's ability to produce narrative macrostructure by looking at the inclusion of story elements, such as the story setting, goals, attempts, outcomes, and internal states of the protagonists. We assessed children's narrative comprehension skills by asking comprehension questions about both a model story that was read to them and about the production story that they generated themselves. Furthermore, we addressed the role of level of intellectual functioning in relation to narrative abilities of children with 22q11DS. Through a comparison with a group of typically developing (TD) children who were younger in chronological age, but similar in terms of their mental age, we investigated whether children with 22q11DS display weaknesses in narrative development that go beyond what may be expected for their level of cognitive development. An additional comparison of the narrative abilities of children with 22q11DS with those of chronologically age-matched children with Developmental Language Disorder (DLD) allowed us to explore whether these two groups of children display comparable narrative difficulties, despite a significantly lower level of intellectual functioning of children with 22q11DS.

4.1. 22q11DS in comparison to TD

Our results showed that children with 22q11DS experience difficulties in both production and comprehension of narrative macrostructure. Regarding the ability to produce narrative macrostructure, we did not detect evidence for a difference between children with 22q11DS and TD children, who were on average more than 2 years younger. Our limited sample size may have prevented us from establishing a difference in the narrative production skills of these two groups. However, if any such a difference exists, our raw data suggests a better performance of the younger TD children. Taken together, our results suggest a delay in the production of narrative macrostructure of children with 22q11DS. This would confirm previous findings regarding the story generation skills of children with 22q11DS (Persson et al., 2006; Van den Heuvel, Reuterskjöld et al., 2017). Furthermore, the absence of a difference with mental-aged matched TD children, who were younger in age, could suggest that in children with 22q11DS, the ability to produce narrative macrostructure is roughly in line with what may be expected given their level of cognitive development. Consequently, we may tentatively infer that the development of narrative production skills in 22q11DS is associated with their level of intellectual

Table A1

Results of the Kendall tau (τ) correlation analyses between narrative comprehension scores on the model and production story on the one hand and IQ, PPVT and CELF scores on the other hand per group.

	22q11DS				DLD				TD			
	Model story		Production story		Model story		Production story		Model story		Production story	
	τ	p	τ	p	τ	p	τ	p	τ	p	τ	p
IQ	−0.25	.251	0.16	.486	−0.08	.725	0.00	1.00	0.36	.134	**−0.47**	**.036**
PPVT	−0.23	.309	−0.03	.900	**0.55**	**.009**	0.13	.564	0.00	1.00	0.00	1.00
CELF	0.05	.816	0.15	.525	0.25	.262	0.05	.814				

Abbreviations. PPVT: Peabody picture vocabulary task. CELF: sentence repetition subtask of the Clinical Evaluation of Language Fundamentals.

functioning.

The present study is the first to examine narrative comprehension in children with 22q11DS. Our findings indicate that children with 22q11DS have more difficulty understanding the production story than the younger TD children. We cannot draw a firm conclusion regarding the comprehension of the model story. However, we believe that our approach may have underestimated the difference between the children with 22q11DS and the TD children, given that the latter group performed at ceiling on the comprehension task of the model story and we observed a medium effect size. This finding may indicate that the level of narrative comprehension of children with 22q11DS is weaker than what is expected based on their cognitive developmental level. In contrast to narrative production, this may imply that the deficit in narrative comprehension in 22q11DS cannot be completely attributed to the overall lower level of intellectual functioning in this population. Plausibly, our finding suggests that other factors besides low intellectual functioning may be contributing to the difficulties in narrative comprehension in 22q11DS.

Taken together, these findings indicate that children with 22q11DS show global weaknesses in narrative skills, with narrative comprehension more affected than narrative production. This is an interesting finding in light of earlier research suggesting that receptive language problems in 22q11DS increase over the course of childhood, leading to a smaller receptive over expressive language advantage in these children in comparison to TD peers (Glaser et al., 2002; Van den Heuvel et al., 2018).

4.2. 22q11DS in comparison to DLD

We did not detect significant differences between the narrative abilities of the children with 22q11DS and the children with DLD, despite the difference in level of intellectual functioning between these groups. Again, while our study would have identified such differences if these were truly substantial in the general population, our limited sample size may have prevented us from detecting smaller group differences. Alternatively, and in line with our expectations, it is likely that we did not detect a difference in narrative performance between children with 22q11DS and children with DLD, because children with DLD often present narrative difficulties irrespective of their level of cognitive functioning (Fey et al., 2004; Norbury et al., 2016; Pearce, James, & McCormack, 2010). Similarly, our observation that the children with 22q11DS did not differ from the children with DLD, despite a lower level of intellectual functioning of children with 22q11DS, may indicate that the difficulties in narrative production and comprehension cannot entirely be attributed to the low level of intellectual functioning in 22q11DS. This is an interesting observation, considering the previously reported overlapping characteristics in the language and behavioral profiles of children with 22q11DS and children with DLD (Goorhuis-Brouwer, Dikkers, Robinson, & Kerstjens-Frederikse, 2003; Swillen, Devriendt, Ghesquière, & Fryns, 2001).

4.3. Possible mechanisms associated with narrative difficulties in 22q11DS

The observed similarities between children with 22q11DS and children with DLD suggest that we can apply our knowledge of mechanisms that are associated with narrative difficulties in DLD to understand narrative difficulties in 22q11DS. Previous studies on children with DLD show that a combination of deficiencies in linguistic, cognitive and social functions may be associated with difficulties in narrative production and comprehension (Blom & Boerma, 2016; Duinmeijer et al., 2012). Linguistically, our participants with 22q11DS demonstrated below average grammatical skills as indicated by their performance on the sentence repetition task. Knowledge of the grammar of a language is important to describe and understand causal connections between story events and episodes (Botting, 2002). Hence, the problems of children with 22q11DS with formulating and understanding grammatical structures may therefore have interfered with their narrative production and comprehension. Furthermore, children with 22q11DS had a similar level of vocabulary comprehension as the younger TD children, which can thus not explain the difference in narrative comprehension between the two groups. However, the understanding of complex sentences and instructions has been reported to be weak in children with 22q11DS (Van den Heuvel et al., 2018). Although we did not measure complex sentence comprehension, it is possible that problems in understanding the sentences that built up the narrative, as well as problems in understanding the questions that were used to assess narrative comprehension may have contributed to the difference in narrative comprehension between children with 22q11DS and younger TD children.

Our findings of the comparisons of narrative comprehension abilities of children with 22q11DS to those of both mental-aged matched TD children and children with DLD are consistent, suggesting that difficulties in narrative comprehension skills cannot entirely be attributed to the weaker intellectual abilities in 22q11DS. With respect to narrative production, our findings are less

straightforward to interpret regarding the role of intellectual functioning, as we did not detect any significant group differences in the comparisons of narrative production abilities. This may indicate that, in reality, children with 22q11DS differ neither from younger mental age-matched TD children nor from children with DLD in their ability to produce narrative macrostructure. Alternatively, differences in narrative production ability may actually exist, but our comparisons failed to demonstrate these at a significant test level, most likely due to *a priori* limited statistical power. Our results suggest that the difference in narrative production between children with 22q11DS and the TD children is larger compared to the difference between children with 22q11DS and children with DLD. Based on these observations, we tentatively speculate that similar to our findings for narrative comprehension, the observed difficulties in narrative production skills in 22q11DS may also not entirely be attributable to a weaker level of intellectual functioning. However, a study based on a larger sample size is required to examine this hypothesis.

In this study, we used IQ scores as a proxy of the level of cognitive functioning of children with 22q11DS. For a deeper understanding of the observed deficits in narrative comprehension as well as narrative production in 22q11DS, it is necessary to examine the narrative skills of children with 22q11DS in relation to broader cognitive abilities, as well as to social functions that have been associated with narrative problems in children with DLD, such as working memory, attention, and the ability to make (social) inferences (Blom & Boerma, 2016; Duinmeijer et al., 2012). In addition, future research with a larger sample could examine the types of errors that children make in the narrative comprehension and production tasks, enhancing our understanding of mechanisms underlying weak narrative functioning.

5. Implications

The deficits in narrative production and comprehension of children with 22q11DS have implications from both a research and a clinical perspective. Many children with 22q11DS experience difficulties in contact with peers and parents, as well as academic problems. Studying narrative difficulties of children with 22q11DS in relation to such functional outcomes is important, given that this may provide an opportunity for intervention. Previous work with low-income preschoolers suggests that narrative intervention may improve children's functioning in school and social settings (Johnston, 2008; Nicolopoulou, Schnabel Cortina, Ilgaz, Brockmeyer Cates, & de Sá, 2015).

Our findings tentatively suggest that the problems in narrative comprehension of children with 22q11DS exceed the severity of problems with narrative production and, moreover, indicate that these comprehension skills are weaker than expected for their level of cognitive development. If future studies replicate this observation, this highlights a challenge for people who interact with children with 22q11DS in a daily life, school or clinical setting with respect to matching their demands and expectations to a child's capabilities (Fiksinski et al., 2018). Namely, the child's relatively stronger production abilities may hide the less readily observable weaker level of comprehension abilities. In addition, the overall level of intellectual functioning may often not reflect the level of language comprehension. Communication partners may overestimate the child's linguistic abilities due to these discrepancies. Therefore, the findings of this study underscore the importance of assessment of language comprehension abilities in children with 22q11DS (see also Van den Heuvel et al., 2018).

6. Conclusion

In summary, we found that children with 22q11DS experience difficulties in their ability to produce narrative macrostructure as well as in their ability to comprehend narratives. Our comparison of children with 22q11DS to younger TD children matched on mental age as well as to age-matched children with DLD, did not allow us to draw a firm conclusion regarding the extent to which narrative production difficulties can be entirely attributed to a low level of intellectual functioning. However, our findings do indicate that difficulties in narrative comprehension of children with 22q11DS were weaker than expected for their developmental level, and may not be solely explained by their overall lower level of intellectual functioning. The relatively weak narrative comprehension skills of children with 22q11DS as compared to their ability to produce a narrative, as well as the potential discrepancy between children's narrative skills and their level of intellectual functioning calls for further consideration from a research as well as a clinical perspective.

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CRediT authorship contribution statement

Iris Selten: Conceptualization, Methodology, Formal analysis, Investigation, Visualization, Writing – original draft. **Tessel Boerma:** Conceptualization, Methodology, Investigation, Writing – original draft, Writing – review & editing. **Emma Everaert:** Conceptualization, Writing – review & editing. **Mariska J. Vansteensel:** Conceptualization, Methodology, Writing – review & editing, Funding acquisition, Project administration. **Jacob Vorstman:** Conceptualization, Methodology, Writing – review & editing. **Frank Wijnen:** Conceptualization, Methodology, Writing – review & editing, Funding acquisition.

Declaration of Competing Interest

None.

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Appendix A

Results of the Kendall tau (τ) correlation analyses between narrative comprehension scores on the model and production story on the one hand and IQ, PPVT and CELF scores on the other hand per group.

We performed a Kendall tau correlation analysis, that is most suited when analyzing associations in a small sample and data measured on an ordinal level. We only explored the relation between children's total IQ score and scores on the language measures on the one hand, and scores on narrative comprehension on the other hand, as we only found a significant group difference on our measures for narrative comprehension. Age did not significantly correlate with the outcomes of these measures of narrative comprehension, and therefore we did not correct for age in our correlation analyses ($\tau < 0.29$, $p > .168$). We report the results in the correlation table below per group (see Table A1). To summarize, we found a significant association between total IQ score and the comprehension score of the production story for the TD children ($\tau = -0.47$, $p = .036$) and a significant association between the PPVT score and the comprehension score of the model story for the children with DLD ($\tau = 0.55$, $p = .009$). We did not find any other significant correlations for TD children ($\tau < 0.36$, $p > .134$), children with 22q11DS ($\tau < 0.24$, $p > .251$) or children with DLD ($\tau < 0.25$, $p > .262$).

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