Emotional Development in the Context of Developmental Disorders

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Abstract

Emotional development is a critically important process that can have major impacts across the lifespan. The current chapter explores what is known about this process in individuals with developmental disabilities, specifically autism spectrum disorder (ASD), fragile X syndrome (FXS), and Down syndrome (DS). It reviews methodological limitations of studying emotional development in developmental disorders and highlights the most prominent and promising methods for use in these special populations. We then systematically review the literature on emotional development in ASD, FXS, and DS with specific focus on recognition and processing of others' emotions, personal expressions of emotions, and emotion regulation. Finally, we discuss implications for treatment and promising future directions.

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Emotion is a core component of what makes us human. Development of emotion is a critical process, which goes from aiding in basic survival early in life to defining social relationships that act as a foundation to most individuals' lives throughout adulthood. There are subgroups of individuals whose emotional processing abilities develop atypically from early in life, or whose atypical development in other domains impacts their early emotional development. Developmental disorders are a broad category that include identified genetic mutations that impact cognitive functioning and behavioral disorders that can impact social functioning while sparing cognitive abilities. Many identified developmental disorders manifest with some impairment in emotional expression, regulation, and processing. These early impairments then go on to impact the social development of these individuals throughout their lives.

It is critical to consider development, which is itself a dynamic process, as playing a crucial role

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in the phenotypic outcomes of individuals with these disorders. Across different disorders, similar profiles of impairment could be due to completely different pathways or developmental cascades (Karmiloff-Smith, 1998). Studying emotional development in the context of developmental disorders presents a promising opportunity to elucidate the underlying mechanisms that are at play in even typical emotional development; in turn, studying atypical development can help explain not only the origins of typical behavior but also the impact of different experiences on the global development of emotion.

While there are many developmental disorders that present with relevant and distinct patterns of emotional development, the current chapter will bring together separate lines of research focusing on autism spectrum disorder, fragile X syndrome, and Down syndrome. We emphasize different levels of analysis that focus on the recognition and interpretation of other people's emotions, patterns of an individual's own emotion expression, as well as emotion regulation abilities across the lifespan. Throughout the chapter, we will highlight physiological measurement and neural processing of emotion in individuals with these developmental disorders and, importantly, will review the methodological challenges in studying emotional development in these populations.

Autism Spectrum Disorder

Autism spectrum disorder (ASD) is a pervasive neurodevelopmental disorder characterized by impaired verbal and nonverbal communication, reciprocal social interaction, and restricted and repetitive behaviors (American Psychiatric Association, 2013). The current estimated prevalence of autism is 1 in 68 children in the United States, with males 4 times more likely to be diagnosed than females (CDC, 2014). Although a singular cause of autism is not known, recent evidence suggests a combination of genetic and environmental factors (Karimi, Kamali, Mousavi, & Karahmadi, 2017; Robinson et al., 2016; Schaaf & Zoghbi, 2011). Autism is a lifelong disorder that differentially impacts those affected, with severity of impairment ranging from mild to severe. DSM-V diagnostic criteria for ASD characterizes persistent deficits in social communication and restricted-repetitive behaviors that are present in early childhood and impact daily functioning, specifying whether symptoms are accompanied by intellectual impairment, language impairment, and associated medical or genetic conditions. Further, based on these deficits, the criteria differentiate severity level of ASD, with level 1 indicating "requiring support," level 2 "requiring substantial support," and level 3 "requiring very substantial support" (American Psychiatric Association, 2013). Thus, the variability in the presence and severity of autism-related symptoms makes the disorder very heterogeneous.

Fragile X Syndrome

Fragile X syndrome (FXS) is a developmental disorder that is associated with high rates of intellectual disability (ID), attention problems, and social anxiety (Hessl et al., 2007; Cordeiro, Ballinger, Hagerman, & Hessl, 2011). FXS also has a high rate of comorbidity with ASD, with many individuals with FXS presenting clinically with restricted and repetitive interests, stereotyped behaviors and impaired social abilities (Abbeduto, McDuffie, & Thurman, 2014; Hagerman & Hagerman, 2001). The most important distinction between ASD and FXS is the identified genetic cause of FXS—an X linked disorder caused by a trinucleotide repeat (CGG) expansion on the fragile X Mental Retardation-1 (*FMR1*) gene on the X chromosome. The full mutation of the disorder occurs when this CGG repeat expansion number is greater than 200 within the 5' UTR region of the *FMR1* gene, at which point the gene becomes methylated, resulting in a loss-of-gene function (Hagerman & Hagerman, 2001). When fully methylated, the gene is silenced and no longer produces its protein product fragile X mental retardation protein (FMRP), which is crucial for proper synaptic functioning and dendritic development (Irwin, Galvez, & Greenough, 2000; Sidorov, Auerbach, & Bear, 2013). The full

mutation of the disorder occurs in approximately 1 in 2500–4000 males and 1 in 7000–8000 females (Hagerman, 2008). The premutation of the disorder, which has a mild cognitive phenotype but puts the person at risk as a carrier for passing the full mutation on to their children, is much more common, affecting between 1 in 500 males and 1 in 200 females (Hunter et al., 2012; Seltzer et al., 2012; Tassone et al., 2012).

Down Syndrome

Like FXS, Down syndrome (DS) is associated with anywhere from mild to severe intellectual disability and has an identified genetic cause. DS is most commonly caused by the presence of a third copy of the 21st chromosome (trisomy 21). This aneuploidy can be partial or complete, and a range of genetic levels of DS exist. The presence of this extra copy of the 21st chromosome leads to an increase in the expression of the protein products of genes located on the 21st chromosome (Chapman & Hesketh, 2000). While there is variability, Down syndrome presents phenotypically with some predictable patterns of physical, biological, and cognitive functioning. The physical phenotype of DS is hallmarked by neonatal hypotonia and identifiable facial and musculoskeletal morphology (Korenberg et al., 1994; Silverman, 2007).

Physical problems associated with DS include, but are not limited to, middle ear disease, problems with the immune and endocrine systems, skeletal and digestive issues, and, most markedly, cardiac defects (Epstein et al., 1991; Chapman et al., 1997). Later in life, there is also a high rate of dementia and a high comorbidity with Alzheimer's disease (Wiseman et al., 2015).

Silverman et al., 2007 reviewed the literature on the cognitive phenotype in DS and found relative weaknesses in expressive language, syntactic processing and verbal working memory, but also found that performance in most cognitive areas could be predicted by individuals' levels of overall intellectual impairment. In a study of young children with DS, Fidler, Hepburn, and Rogers (2006) found that socialization abilities were the only area that set the toddlers with DS apart from a mixed developmental disabilities group. This shows that even early in life, sociability seems to be a relative strength in DS (Fidler et al., 2005; Fidler et al., 2006).

DS and FXS, while not the most prevalent developmental disorders, are good examples of genetic disorders who present clinically with differing patterns of social and emotional abilities. These disorders can each provide a rich context in which to study the differential impact a genetic insult can have on emotional dysregulation. Alternatively, highlighting ASD, which does not have a single, identified genetic cause, but does present with marked social impairments, allows us to investigate the impacts of social impairment on emotional development.

This chapter will highlight both what is known about, and the gaps in our understanding of, emotion development in these three disorders. It will emphasize the impact these disordered developmental trajectories can have on the development of emotional processing and suggest ways in which this knowledge can facilitate the development and implementation of evidence-based treatments. Importantly, it explores the premise that investigating emotional processing in children with these developmental disorders can shed light on the underlying genetic, neural, and behavioral mechanisms that are at play in the emotional development of children who are typically developing.

Methodologies for Studying Emotion in the Context of Developmental Disorders

Before reviewing the literature on emotional development in these developmental disorders, it is important to note that some common methodologies used to study the development of emotion cannot feasibly be utilized in individuals with developmental disorders.

Parent report (typically via the completion of questionnaires) is a common method used to investigate the emotional development of the child. Parent report data is inherently limited,

given the second-hand nature of the information, as well as the very personal nature of the experience of emotion (Lagattuta, Sayfan, & Bamford, 2012). This limitation can be amplified when the child has a developmental disorder that makes it difficult for him or her to communicate their emotional experiences either due to reduced language ability or reduced emotional awareness. While some studies show that change over time in emotion expression or response to treatment can be reliably captured by parent report in children with developmental disorders (Hagerman et al., 2016), it is important to consider that in disorders such as FXS and ASD, the parents themselves have been shown to exhibit heightened levels of anxiety and depression (Cohrs & Leslie, 2017; Cordeiro et al., 2011), and those differences could influence the ratings of their children. Another large limitation of using standardized, parent-report measures is the fact that these measures have been normed on typically developing populations, so there is often a problem with floor or ceiling effects (Hessl et al., 2008).

Behavioral methods that require an overt response are often used to measure attention or reaction to emotion in typically developing children. When using these methods with children with developmental disorders, however, researchers must be careful to design paradigms that are not too cognitively challenging, and that will not overstimulate or overtax the participants.

A common problem is the interference of one area of deficit in the attempted measurement of another. For example, when asking a child with FXS about their hypothetical emotional response to a given situation, their social anxiety, attentional problems, or ability to think hypothetically may limit or alter their response, and the resulting data may reflect those areas of impairment rather than a true representation of that child's understanding of their own emotional responses.

Functional neuroimaging methods like functional magnetic resonance imaging (fMRI) are commonly used across development in a diverse number of developmental disorders. fMRI methods allow researchers to ask questions about the neural circuitry that is affected in a given disorder, and where emotional processing is concerned, the relevant structures are often medial temporal lobe areas such as amygdala, and orbitofrontal cortex. In disorders like FXS, we can investigate the direct impact that atypical genetic functioning can have on brain development and function (Hessl, Rivera, & Reiss, 2004; Kim et al., 2012; Rivera, Menon, White, Glaser, & Reiss, 2002). This can provide insight into the underlying causes of atypical aspects of emotional development and can give researchers specific neural targets for measuring efficacy of treatment efforts. Most functional neuroimaging methods require relatively significant levels of compliance, including the need for participants to follow instructions and stay still, which can be prohibitive for use in individuals with intellectual disability (ID). This often precludes use of these neuroimaging methods in populations with ID, or limits the applicability of findings to the whole disorder when only the highest cognitively functioning individuals can be included.

Event-related potentials (ERPs) have been used in populations that have less success laying supine in an MRI environment, though tolerance of the cap placement can still be challenging and the need for stillness remains a factor, though to a lesser degree than with fMRI. ERPs also have the advantage of providing an environment less likely to cause sensory over-stimulation (compared to loud, confined spaces of the MRI environment). This is an important factor given the high rate of sensory sensitivities in some populations with developmental disorders (Rogers, Hepburn, & Wehner, 2003). Passive viewing ERP paradigms (rather than those requiring instruction-following and overt motor responses like a button press) can be very helpful in studying infants and young children with developmental disorders and can thus help reveal answers to many of the interesting questions about early emotional development. While much neuroimaging research focusing on emotion investigates functioning and connectivity of the amygdala, ERP methodology cannot directly measure brain activity coming from such a subcortical structure. While this is a limitation, a substantial literature exists looking at attention-related components

that can help us understand emotional processing across development (Hajcak, MacNamara, & Olvet, 2010 for review).

Eye tracking is one very useful methodology when investigating emotional processing and can be effectively implemented across development. Eye tracking can be used in populations from very young infants to adults, and tasks that are designed to allow for passive viewing can be administered to individuals with even severe ID. Eye tracking allows for researchers to tap into attentional processes that influence emotional development across the lifespan. The method can be used to investigate many critical questions as to the mechanisms and origins of patterns of emotional development in different populations with developmental disorders.

Given the nuanced nature of testing emotional development, care must be taken in choosing and evaluating the appropriate methodology for use with different populations. The following sections of the chapter will highlight findings in ASD, FXS, and DS utilizing several methods across a wide age range of individuals. We will highlight the methodological shortcomings and modifications that were made to allow for use in these special populations.

Emotional Development in Autism Spectrum Disorder

It is broadly understood that a hallmark of ASD is disordered social-emotional processing, particularly recognizing and understanding the reciprocity of emotions in both verbal and nonverbal social interactions (Bons et al., 2013). Difficulties in processing of emotion, such as identifying and describing feelings, distinguishing bodily sensations of emotional arousal, attention to the eyes for social information, and facial expressions of emotion have been well-documented in individuals with ASD (Bons et al., 2013; Hill, Berthoz, & Frith, 2004). Some researchers have even suggested that individuals with ASD suffer from "mindblindness", or an inability to interpret others' mental states (Baron-Cohen, 1997). These observations suggest deficits in the social domain for individuals with ASD and may be a result of the inability to properly process a broad spectrum of emotional information (Philip et al., 2010).

It has been hypothesized that the hallmark social impairments seen in ASD may be consequences of an abnormally functioning "mirror neuron system" (Dapretto et al., 2006; Decety & Moriguchi, 2007; Williams, Whiten, Suddendorf, & Perrett, 2001). The findings in the literature on mirror neuron dysfunction in ASD are mixed, with some neuroimaging studies indicating a deficit in mirror neuron function in ASD (Hadjikhani, Joseph, Snyder, & Tager-Flusberg, 2006; Nishitani, Avikainen, & Hari, 2004), while others report no marked differences in the mirror neuron system of individuals with ASD (Fan, Decety, Yang, Liu, & Cheng, 2010; Pokorny et al., 2015; Pokorny, Hatt, Rogers, & Rivera, 2017; see Hamilton, 2013 for review).

Individuals with ASD are sometimes thought to lack empathy (Baron-Cohen & Wheelwright, 2004); however, Smith (2009) clarified that while individuals with ASD may have weak *cognitive empathy*, many appear to have intact *emotional empathy*; i.e., the ability to ascertain another individual's emotions and respond with similar emotion. The literature is decidedly mixed, with some studies showing a typical level of emotional empathy in high-functioning individuals with ASD (Dziobek et al., 2008), and others showing an impaired emotional empathy ability (Baron-Cohen, 2002; Minio-Paluello, Baron-Cohen, Avenanti, Walsh, & Aglioti, 2009; Williams et al., 2001). Alexithymia is a personality construct that is defined by an inability to identify and explain one's own emotional state (Bird & Cook, 2013). The literature is unclear as to the connection between alexithymia and ASD, though some have posited that alexithymia may be a co-occurring factor rather than a feature of autism, given its presence in other, unrelated disorders (Bird & Cook, 2013).

Very early in development, infants later diagnosed with ASD exhibit diminished eye contact, difficulties with joint attention, decreased social smiling, and orienting to their name (Osterling & Dawson, 1994; Ozonoff et al., 2010). As such, face processing in autism has been intensely

studied as an early indicator of downstream atypical social cognition. Studies of face processing in autism yield mixed evidence, but overall present a pattern of atypical processing of facial information present early in infancy and persisting into adulthood. Various eye tracking studies have suggested altered visual scanning of the face, with reduced time spent looking at the eyes and overall core features of the face (Chawarska & Shic, 2009; de Wit, Falck-Ytter, & von Hofsten, 2008; Jones, Carr, & Klin, 2008; Jones & Klin, 2013; Klin, Jones, Schultz, Volkmar, & Cohen, 2002; Pelphrey et al., 2002). Across studies, meta-analyses report impairments in gaze fixation to the eyes and reduced attention to social information (Papagiannopoulou, Chitty, Hermens, Hickie, & Lagopoulos, 2014); however, mixed findings of intact face scanning and social orienting emphasize the need for further investigation (Guillon, Hadjikhani, Baduel, & Rogé, 2014).

ERP studies investigating the neural processing of faces report atypical face perception in autism. Specifically, numerous studies have investigated the N170 ERP component, a neural marker of face processing, and report delayed N170 latencies that reflect slowed processing (Batty, Meaux, Wittemeyer, Rogé, & Taylor, 2011; Kang et al., 2017; McPartland, Dawson, Webb, Panagiotides, & Carver, 2004; Stavropoulos, Viktorinova, Naples, Foss-Feig, & McPartland, 2018; Webb, Dawson, Bernier, & Panagiotides, 2006). A recent fMRI meta-analysis investigating the neural correlates of emotional face processing in autism showed atypical activation in subcortical structures implicated in face processing, including the amygdala, hypothalamus, and basal ganglia (Aoki, Cortese, & Tansella, 2015). Moreover, findings report under-connectivity between the fusiform gyrus and visual cortex in individuals with autism during a face recognition task (Lynn et al., 2018). Together, this research suggests that atypical activation in subcortical structures may underlie altered perceptual encoding of faces in individuals with autism.

In addition to the documented differences in *interpreting* others' emotions, atypical *expression* of emotions has also been observed in ASD. Individuals with ASD show impaired motor empathy, or facial mimicry abilities across multiple emotions: happy, sad, fear, anger, disgust, and surprise (Bons et al., 2013). Also, Brewer et al. (2016) found the emotional expressions produced by individuals with ASD are not perceived as well as those produced by typically developing controls; interestingly, by both controls and other individuals with ASD (Brewer et al., 2016). Begeer, Koot, Rieffe, Terwogt, and Stegge (2008) showed that development plays a strong role in emotion expression, evidenced by data showing that young infants who go on to be diagnosed with ASD show emotions in a similar way to TD controls, but as age increases, their expressions tend to become less spontaneous and less socially oriented.

Emotional outbursts and tantrums can be very common in young children with ASD (Konst, Matson, & Turygin, 2013; Maskey, Warnell, Parr, Le Couteur, & McConachie, 2013; Mazefsky, Pelphrey, & Dahl, 2012; Samson, Hardan, Podell, Phillips, & Gross, 2015). Though the more traditional social and communication problems are what often lead to a child receiving an ASD diagnosis (Dawson, 2008; Dawson et al., 2010), emotional dysregulation (i.e., increased negative affect, or feelings of emotional distress and decreased positive affect) is observed and retrospectively reported by parents of children with ASD (Garon et al., 2009; Ozonoff, Williams, & Landa, 2005; Wimpory, Hobson, Williams, & Nash, 2000). These dysregulated patterns of affect continue throughout development (Mazefsky et al., 2012; White, Oswald, Ollendick, & Scahill, 2009).

Emotional and behavioral difficulties commonly seen in autism may be explained by underlying deficits in emotion regulation (ER), a process by which one regulates their own emotions behaviorally, cognitively, and physiologically (Berkovits, Eisenhower, & Blacher, 2017; Gross & Jazaieri, 2014; Mazefsky et al., 2013). Maladaptive emotion regulation skills, or emotion dysregulation, in ASD is associated with behavioral disturbances such as uncontrollable outbursts or aggression (Mazefsky & White,

2014) and have implications for anxiety and depression (Mennin, Holaway, Fresco, Moore, & Heimberg, 2007; Weiss, Thomson, & Chan, 2014). Cognitive reappraisal, an antecedent-focused regulation strategy, can be used to down-regulate negative emotional responses and reframe the situation to decrease emotional responsivity (Gross, 1998). Importantly, the ability to use cognitive reappraisal is associated with positive outcomes such as reduced mood and anxiety problems, sense of purpose in life, personal growth, and better interpersonal functioning (Gross & John, 2003). Behavioral evidence suggests that children with ASD employ more maladaptive coping strategies (e.g., increased venting, avoidance, diminished problem solving, increased resignation from task) in frustrating situations (Jahromi, Meek, & Ober-Reynolds, 2012; Konstantareas & Stewart, 2006). Emotion dysregulation has been reported to remain stable throughout childhood in ASD, with declines in social skills and atypical coping strategies that contribute to increasing internalizing and externalizing behaviors (Berkovits et al., 2017; Rieffe et al., 2011). Adolescents with ASD report involuntary employment of maladaptive ER strategies, including rumination, increased emotional arousal, and disengagement (numbing and inaction) (Mazefsky, Borue, Day, & Minshew, 2014). Further, adolescents with ASD show less frequent use of cognitive reappraisal (even when prompted to use this strategy) and instead expressive suppression (Samson et al., 2015), a pattern that continues into adulthood (Samson, Huber, & Gross, 2012). Together, these studies highlight problems in emotion regulation, present early in life and continuing throughout development.

Although multiple studies have identified emotion regulation problems in autism, further research is needed to delineate origins of these problems. To date, only a small number of studies have directly investigated the neural and physiological features of ER in autism. Further, the literature suggesting a disrupted autonomic nervous system (ANS) related to emotion regulation and social functioning in autism has yielded mixed results (see Benevides & Lane, 2015 for review). Several studies report a relationship between reduced respiratory sinus arrhythmia (RSA) amplitude and disrupted ER, internalizing/externalizing behaviors, and anxiety (Bal et al., 2010; Guy, Souders, Bradstreet, DeLussey, & Herrington, 2014; Neuhaus, Bernier, & Beauchaine, 2014). However, in a recent study of young children with ASD, researchers measured heart rate variability and ER strategies during a frustration-eliciting task. Despite difficulties in employing effective coping strategies, the underlying physiological arousal (heart rate) of emotion was intact, and differences emerged only in behavioral and expressive stages of ER (Zantinge, van Rijn, Stockmann, & Swaab, 2017).

Neuroimaging studies report abnormal prefrontal cortex (PFC)-amygdala connectivity in autism, suggesting that this contributes to problems with emotion regulation and anxiety (Swartz, Wiggins, Carrasco, Lord, & Monk, 2013). A preliminary fMRI study explored the neural mechanisms of cognitive reappraisal of disgust in children and adolescents with ASD (Pitskel, Bolling, Kaiser, Pelphrey, & Crowley, 2014). Researchers reported that although participants with ASD were able to behaviorally modulate their emotional response to disgust, they exhibited atypical neural modulation of insula and amygdala, and decreased connectivity between amygdala and prefrontal cortex (Pitskel et al., 2014). Consistent with these findings, in a study of cognitive reappraisal of faces, adults with autism showed altered activation in the nucleus accumbens, amygdala, and dPFC (Richey et al., 2015). Taken together, these studies highlight the importance of autonomic reactivity and brain connectivity associations in emotion regulation, and suggest mechanisms of disrupted emotion regulation in autism.

While on the one hand the presence of disrupted emotional development in ASD is well-documented, there are nonetheless many unanswered questions, and the literature is still mixed with regard to findings of dysfunction. Many of these contradictions in the literature may be related to the myriad of different methodological approaches to studying emotional development in ASD. It addition, ASD is inherently variable and heterogeneous, so characterizing the

entire population is not only challenging, but perhaps the incorrect level of analysis. Because of this heterogeneity, there are many studies that are aimed at subtyping individuals with ASD based on similar patterns in development (Amaral et al., 2017; Singer, 2005).

Moving forward in the study of emotional development in ASD, more studies are needed that both identify distinct emotional profiles of individuals with ASD and target the mechanisms underlying the differences seen. While we know a great deal about the behavioral manifestation of emotion dysregulation in ASD, we don't yet have a firm grasp on what is driving these differences, biologically. In the future, a more holistic approach should be taken, in which the larger context of development, including environmental impacts and the impact that cognitive factors have on emotion regulation abilities in ASD are taken into consideration. For example, it is important to consider how language or executive functioning skills (two areas of cognition that are variably impacted by ASD) impact emotional processing and regulation abilities.

Emotional Development in Fragile X Syndrome

Before exploring the literature on emotional development in FXS, it is necessary to discuss some of the challenges that exist in studying this disorder, and others with similar phenotypes, which may influence what can be known and measured in the population. Due to the X-linked nature of FXS, a large proportion of the literature focuses on only males, limiting the applicability of findings to the whole population. Given that in the majority of males with FXS the gene is thought to be methylated/silenced, focusing on males also inherently limits the amount of information we can glean about the developmental impacts of variable levels of FMRP, the protein product of the *FMR1* gene.

Another challenge in studying emotional development of children with FXS is the impact that their intellectual disability can have on their ability to participate in research studies and the way that cognitive ability may interfere with the way that we measure emotional processing. It would be unfortunate to conclude that individuals with FXS have a deficit in an area in emotional development, if in fact the deficit was due to their inability to follow the complex instructions of the task. This also highlights the need for appropriate control groups, matched on important factors such as intellectual ability, which is an area of great concern when studying groups of individuals with developmental disorders (Karmiloff-Smith, 2009).

While the FXS literature is much smaller than that of ASD, there are still a number of studies that have investigated how emotional development occurs in the context of this single gene disorder. These findings help us not only learn about how emotional development can go awry but also learn much about the underlying mechanisms that fuel emotional development in general.

One of the main behavioral challenges in FXS is the extremely heightened level of anxiety seen in this population. Cordeiro et al. (2011) reported that up to 82.5% of individuals with FXS ages 5–35 years qualified as having clinical levels of anxiety. Social phobia was more common in adults than children with FXS, but social phobia and specific phobia were the most common across development, with 58% of the sample qualifying for multiple anxiety disorder diagnoses (Cordeiro et al., 2011). Young children with FXS who do not have anxiety disorders show less attention problems, hyperactivity, and aggression than those with anxiety or those with heightened anxiety and ASD (Talisa, Boyle, Crafa, & Kaufmann, 2014). It is important to keep in mind that atypical emotional processing, including processing of other's emotions and regulating one's own emotions, may be underlying much of this anxiety.

Simon and Finucane (1996) found that adult males with FXS showed no evidence of a deficit in ability to identify emotional facial expressions. Bouras, Turk, and Cornish (1998) also found no evidence that young boys with FXS as a group have an impaired ability to recognize the expression of basic emotions. While it is widely accepted that children with FXS have social impairment, this impairment does not

seem to include or result from an impairment in either facial identification or in the basic perception of facial emotion.

Farzin, Rivera, and Hessl (2009) showed children calm, happy, fearful, and scrambled faces and found that individuals with FXS made fewer fixations to the eyes than typically developing individuals, but interestingly only for real faces (not for the scrambled face), showing that the effect was indeed face-specific. These findings are consistent with previous findings showing children, adolescents, and adults with FXS having greater avoidance of eye contact in social interactions globally (Einfeld, Tonge, & Florio, 1994). Individuals with FXS also showed greater pupillary responses to emotional faces than controls, hinting at a processing difference at the neurophysiological level (Farzin et al., 2009). Relatedly, Ballinger, Cordeiro, Chavez, Hagerman, and Hessl (2014) showed that individuals with FXS showed significantly reduced startle potentiation to fearful faces than the typically developing control group. The authors interpreted these findings as indicating differential amygdalar responsiveness to social stimuli as a contributing factor to phenotypic variability among individuals with FXS.

Kim et al. (2012) also showed atypical amygdala response in adolescents on the FX spectrum. The study showed participants neutral, happy and fearful faces in an fMRI paradigm. Results revealed an expected overall increase in amygdala activation to emotional faces, but a blunted response to fearful faces, specifically. This differed from the heightened response to fearful faces that was seen in the typically developing control participants. Furthermore, the degree of blunting of this response was directly correlated with both gene expression and anxiety level, with the most anxious and the most genetically impacted individuals showing the greatest degree of atypical amygdala response. A separate fMRI study confirmed that, while emotion recognition is relatively intact in FXS, the brain circuit responsible for such processing, and for modulating responses to emotional faces may be functioning atypically (Hagan, Hoeft, Mackey, Mobbs, & Reiss, 2008).

In a recent study, Burris et al. presented one of the first studies to investigate attention to emotional faces in young children and infants with FXS (Burris et al., 2017). It was found that when presented with emotional faces in the context of a dot probe task (a task designed to quantify implicit attentional biases) presented on an eye tracker, infants and young children with FXS showed a threat-specific attentional bias. These results suggest that the attentional systems of these young children are preferentially vigilant to detect fearful facial emotions compared to neutral faces, and more so than to happy emotions. This study indicates that, even in infancy, there is a basic difference between the way the brains of individuals with FXS are processing and reacting to fearful emotional facial displays of others.

There is some research addressing the environmental factors that underlie the emotional differences in FXS. In the context of a demanding environment, individuals with FXS are commonly unable to emotionally regulate and sometimes turn to self-injurious behavior (Hall, Lightbody, & Reiss, 2008; Symons, Clark, Hatton, Skinner, & Bailey, 2003). van Lieshout, De Meyer, Curfs, and Fryns (1998) found that some environmental factors, specifically parental anger, was negatively correlated with the emotional stability of children with FXS. Hessl et al. (2001) also reported that levels of parental psychopathology were predictive of internalizing and externalizing problems in young children with FXS. They also linked the amount of FMRP in girls with FXS to heightened levels of social withdrawal and anxious and depressed behavior. Much of the literature points to basic differences in the processing of emotions at a neural level across development in FXS, but there is evidence suggesting that environmental factors, such as parenting and maternal sensitivity, may impact emotional development in this population as well (Hauser, Kover, & Abbeduto, 2014; Smith, Hong, Greenberg, & Mailick, 2016). Importantly, these differences in neural functioning and outcomes related to environmental factors have both been directly linked to the genetic output of the impacted gene in FXS.

While much is known regarding the genetics and neural impacts, there are still many important unanswered questions about emotional development in FXS. Burris et al., 2017 demonstrated that individuals with FXS have a threat-specific bias in their attention, something that in other populations has been indicated as a risk factor for anxiety, yet we do not yet know if these biases are directly linked to anxiety levels in older individuals with FXS. Making this concrete connection could help elucidate the neural mechanism underlying emotion dysregulation in FXS and, in doing so, also shed light on the underlying mechanism that exists in the absence of this single gene disorder. In addition, understanding the impact of this gene mutation on the development of emotional attention could open doors to targeted treatment and further understanding about the genetic factors contributing to social anxiety.

Emotional Development in Down Syndrome

The literature on emotion development in DS is much smaller than that of ASD, perhaps because a large amount of the research in DS focuses on investigating the molecular genetic component of the disorder rather than the behavioral. There is, however, some work investigating emotional processing abilities of individuals with DS, and how these skills develop and change across the lifespan.

Carvajal and Iglesias (2002) reviewed the literature and found that children with and without DS present with similar patterns of emotional development when measured in terms of face-to-face interaction between mother and infant. The small differences found were attributed to difference rooted in the DS population's impairments in inhibitory control. These findings highlight the important impact that level of intellectual disability may have on patterns of emotional development.

There are mixed findings in the literature focusing on face processing in DS. Annaz, Karmiloff-Smith, Johnson, and Thomas (2009) found that when compared with both high- and low-functioning groups of children with ASD and children with Williams syndrome, children with DS processed holistic faces better than isolated features of faces. They outperformed all other groups in recognizing an upright whole face. In adults with DS, emotional face processing was impacted by perseverative errors to the lower half of the face, a pattern not shown by a group of intellectually disabled peers (Carvajal, Fernández-Alcaraz, Rueda, & Sarrión, 2012). Kasari, Freeman, and Hughes (2001) showed that children with DS can correctly identify emotional facial expressions, but not at the level of chronologically age-matched peers, indicating a present but impaired skill. Interestingly, when mistakes were made by children with DS, they were most likely mislabeling negative emotions as positive emotions. Unsurprisingly, the young children with DS in this study struggled to verbally label the emotions and struggled as the difficulty of the task increased. In a 2-year follow-up of this study sample, it was shown that the participants with DS showed no change in abilities to identify or recognize emotions as they aged. Porter, Coltheart, and Langdon (2007) saw a similar impairment in identification and labeling of emotion, but only for negative emotions, with the group again often labeling negative emotions as positive, and exhibiting specific difficulty when labeling sadness. Similarly, children with DS struggle to match surprise and fearful facial expressions (Wishart & Pitcairn, 2002; Williams, Wishart, Pitcairn, & Willis, 2005).

When it comes to individuals' experience or expression of emotion, individuals with DS present with a distinct pattern when compared to those who are typically developing and those with other developmental disorders or intellectual impairment. Evidence suggests that children with DS may show more positive emotional signals overall than other children with ID. Fidler and Barrett (2006) showed that children with DS smiled more frequently than other groups of children with ID. Interestingly these results changed as individuals with DS aged, with no difference being shown by adulthood. It has been hypothesized that children with DS may tend to rely heavily on their positive emotional responses

and social skills to compensate for their weaker areas of cognitive functioning (Freeman & Kasari, 2002).

In a study investigating expression of empathy in school-aged children with DS, Kasari, Freeman, and Bass (2003) found that children with DS demonstrated prosocial behavior when shown an experimenter in distress but failed to react empathically when shown a socioemotional vignette featuring different emotions. This shows that children with DS can show an empathic emotional response to others, but that this skill may be context dependent. Very little research has been done on emotional regulation in DS, though Bieberich and Morgan (2004) showed that when compared to ASD peers, individuals with DS show more stable levels of self-regulation over time and present greater positive affect stability, overall.

What We Can Learn About Emotion Development from Studying Developmental Disorders?

Studying emotional development within atypically developing populations of children presents a promising opportunity to build a framework in which we can track the cascade of underlying processes that influence emotional development in the general population. By looking at an identified developmental disorder, we can think of this developmental cascade as starting with atypical neural circuitry which can lead to altered physiological and biological responses that then relate to deficits in executive functions like global attention, cognitive control, and cognitive inhibition. In turn, these conditions can impact patterns of face processing that can lead to downstream deficits in emotion regulation and recognition of emotion, which eventually may lead to impairments in social and communication skills. Thus, using atypical development as a model, we can work backward up the developmental cascade to attain a greater understanding of which factors are most impactful in typical emotional development. Based on the classic principles of multifinality, we know that there are multiple pathways to get to the same phenotypic outcome in development. Utilizing patterns of emotional development shown in developmental disorders can help us highlight the impact that these specific divergent pathways can have on emotional outcomes (Cicchetti & Rogosch, 1996).

Furthermore, looking at emotional development through the lens of atypical development allows us to take a cognitive neuroscience approach, given that the perturbations we see in these disorders allow us to investigate specialization of brain regions and their respective functions. Indeed, as lesion studies have well demonstrated, working backwards from insults in development can allow us to reach conclusions that would not have been possible without these models of atypical development.

Within this developmental cascade and neuroscience framework, there are numerous domains in which we can highlight the importance of studying atypical development. Utilizing developmental disorders, we can focus on developmental timing and the role that global development has on emotional development specifically. One example of developmental timing that can be aided by the investigation of atypical development is sensitive and critical periods (Johnson, 2005). Developmental disorders can provide great insight into the timing and flexibility of these crucial periods in development and how development itself, along with individual experiences, come together to influence outcomes (Karmiloff-Smith, 2018). Studies within this framework often focus on sensory domains in developmental disorders, but we argue here that great insights can also be gained from studying emotional development in developmental disorders.

There is also much to be learned from examining the role of individual differences across groups of children who are both atypically and typically developing. Often in research focusing on typical development, there is a false assumption of homogeneity. In the literature of developmental disorders, we see that there are many factors that can impact outcomes, and that these factors may differ across individuals. As an example, fragile X syndrome allows us to

investigate a titrated contribution of the *FMR1* gene, and evidence has shown that individual levels of involvement of this gene are linked with behavioral, psychophysiological and neural outcomes (Kim et al., 2012).

Studying emotional development in developmental disorders also affords us an opportunity to investigate compensatory mechanisms early in life, and the impact of neural plasticity, which can be an important tool for the development of interventions, but also helps us learn about different pathways that could be in place in typical emotional development.

Unanswered Questions and Future Directions

Arguably, the most important unanswered question in the literature on these developmental disorders is that of how to treat emotional dysregulation and anxiety in these populations. Before evidence-based and individually tailored treatments can be developed, however, there are still many questions regarding the basic, biological mechanisms underlying these deficits and, critically, how these mechanisms develop over the lifespan that have yet to be answered.

One such avenue for treatment of emotional problems in disorders (such as FXS) that presents with a specific attentional bias toward threat is attention bias modification (ABM) (Amir, Beard, Burns, & Bomyea, 2009; Bar-Haim, 2010; Hakamata et al., 2010). ABM treatment focuses on systematically training individuals who have an attention bias toward threat to either attend more toward positive or neutral stimuli instead of focusing on threatening stimulus. This type of attention training paradigm could be a prime candidate for use with a population with intellectual disability, given that it can be passive viewing. A 2017 review highlights the technique as a promising new avenue to treat the emotional underpinnings of anxiety but cautions against overinterpretation of the literature, given the small effect sizes and replication failures (Mogg, Waters, & Bradley, 2017).

There are many behavioral therapies that have been utilized to treat ASD in young children, ranging from after-school social skills training groups to intensive one-on-one therapy with a clinician. While an emotional component is incorporated into most of these behavioral interventions, some of these treatments highlight this domain more than others. A primary goal of the Early Start Denver Model (ESDM) is to target deficits in socio-emotional and communication domains that are impacted in ASD. Emotion sharing is one of the key domains of ESDM (Rogers & Dawson, 2010) and as such, ESDM treatment focuses heavily on fostering greater eye contact and facial gaze, which could improve emotional facial expression processing and encourage emotional mirroring.

Computer game-based interventions that focus on social skills training and emotion recognition training are also gaining popularity. In a 2017 review, Grossard et al. identified 31 such game-based interventions that target teaching social skills and emotion recognition. While there are some aspects of these therapeutic games that are encouraging, such as their appeal to the targeted population, there are still many shortcomings. In the majority of cases, these methods are still targeting only high-functioning individuals, and they still tend to not meet standards of treatment efficacy required for clinical trials. Thus, while therapeutic game-based techniques are promising, they still have a way to go before being applicable across the full spectrum of individuals with developmental disorders that exhibit atypical emotional development.

It is clear from the literature that developmental disorders like ASD, FXS, and DS come with varied profiles of atypical emotional development. These atypical patterns include components of emotional processing from interpreting and reacting to others' emotions and expression and regulation of one's own emotions and can be identified at the behavioral, physiological, neural, and genetic levels. They clearly impact the phenotype of these disorders, and in some cases, are defining components in their presentation. Many developmental factors play a role in these atypical patterns of emotion, with some studies

documenting change over time, and others pointing to individual differences related to parenting factors, environmental factors and cognition. Future research directions should therefore focus on studies that include a range of ages and phenotypic presentations, and should employ methods that both tap into underlying biological processes and that can be used across a range of cognitive abilities.

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