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Chapter 31

Atypical Sense of Self in Autism Spectrum Disorders: A Neuro-Cognitive Perspective

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1. Introduction

1.1. Self – Definitions and concepts

The concept of self is notoriously difficult to define and different notions and theories of the self have been proposed by a variety of disciplines all interpreting concepts of self and identity in various ways. We adopt the definition advanced by neuroscientists Kircher and David [1] who interpret the self as ‘the commonly shared experience, that we know we are the same person across time, that we are the author of our thoughts/actions, and that we are distinct from the environment’ (p.2). In cognitive neuroscience literature, operational definitions of the self are used which are measurable by experimental methods including self recognition, self and other differentiation, body awareness, awareness of other minds, awareness of self as expressed in language and important concepts such as autobiographical memory and self narrative. There is significant interest in the role of the self and possible abnormalities associated with the self, as causally implicated in autism. In this paper we review developmental perspectives of self and self-related functions with reference to their neuroanatomical basis and investigate the possible causes for atypical self-development in Autism Spectrum Disorders (ASD).

2. Review of studies investigating the self in ASD

The question whether individuals with ASD have a different sense of self than people without ASD, i.e. ‘neurotypicals’ has fascinated researchers and clinicians for decades. Kanner [2] in 1943 already noted self-deficits in the children he described, including their difficulties in maintaining a constant self-concept, and associated problems of adapting their fragile

3. Atypical development of self-related processes in ASD

There is general consensus among developmental theorists [e.g. 10,11] that early interpersonal communication is central to the establishment of the self in normal development. A large body of research literature indicates that early processes underlying self-other awareness are impaired or delayed in autism including gaze following, abnormal response to sound and deficits in attention [12], showing of objects, responding and orienting to own name [13], looking at other’s faces [14], pretend play, protodeclarative pointing and gaze monitoring [15], empathy and imitation [16], joint-attention behaviour [17], affect and personal relatedness [18,19].

3.1. Self recognition and awareness

The ability to recognize one’s own face in the mirror is considered a test for ‘self-awareness’. Self-recognition as measured by mirror tests [20] in 18 months old children has been depicted as a developmental milestone in self-conception and described as the ‘achievement of a cognitive self’ [21]. Not only is self recognition essential for developing a concept of self, and self-other differentiation, it is also a prerequisite for later developing theory of mind abilities, as a stable self concept is the basis for being able to read the mental states of others. Research data on self-recognition reveals that autistic children’s responses to their mirror images are qualitatively different from those of normal children [22]. Children with ASD have deficits of self-awareness as measured by a self-recognition test [23] they show little interest in their own mirror images and have been described as relatively ‘face inexperienced’ [24].

The prefrontal cortex, especially the Right Hemisphere (RH) plays a critical role in the recognition of one’s own face [25] as evidenced by functional imaging studies. Face perception studies in individuals with ASD suggest abnormal functioning in the fusiform
face area as well as amygdala, brain regions associated with the ‘social brain’ (e.g. 26,27) involving the RH.

3.2. Self/other differentiation

The ability to differentiate between self and other is also essential for the development of self-awareness, which appears to be impaired in autism. In particular, the recognition of a separate existence of other people seems to be delayed in children with autism [28,29]. Attentional abnormalities, such as ‘tunnel vision’, the tendency to think in a monotropic manner, have been suggested by some as the cause of ‘self-other problems’ in ASD [30]. Donna Williams [31] interprets monoprocessing as the inability to process simultaneously information of oneself and others.

In neurotypicals the middle cingulate cortex and ventromedial prefrontal cortex are involved in self/other processing. In contrast, atypical neural responses have been reported in individuals with ASD. A recent fMRI study [32] investigating the attribution of behavioural outcomes to either oneself or others while playing an interactive trust game revealed a lack of brain activity in the cingulate cortex indicating diminished ‘self responses’ in individuals with ASD. However, ‘other responses’, attributing actions to other people were intact. Previous research data using trust games [33] had demonstrated that cingulate cortex activation is consistent with self-response patterns generated during interpersonal exchanges. Chiu et al. [34] interpreted their data in terms of a deficit in ASD in monitoring their own intentions in social interactions and thus contributing to impaired theory of mind abilities, lack of introspection and self-referential processing. Of particular interest is the fact that the ‘impaired self-responses’ in the ASD group correlated with their behavioural symptom severity, i.e. the lesser activity along cingulated cortex the more serious were their behavioural symptoms. Similar results have been reported by Lombardo et al. [35] also demonstrating atypical neural responses from the middle cingulate cortex during a self-referential processing task. This study also provided a link between these deficits and early social impairments in autism. In addition, these authors also demonstrated reduced functional connectivity between ventromedial prefrontal cortex and lower level regions (e.g. somatosensory cortex) in individuals with ASD during these self-representation tasks.

Previous studies identified the right inferior parietal lobe, along with frontopolar and somatosensory regions [36,37] as critical for distinguishing between self and other. Additional data [e.g. 38] demonstrate that SI and SII cortices, which contribute to the mirror-neuron system, are also crucial for preserving a sense of self.

3.3. Body awareness, sense of agency

Knowing oneself and knowing one’s body are closely related concepts. In his review on body image and the self, Goldenberg [39] argues that the acquisition of body image is not innate but acquired through experiences of one’s own and other bodies. Likewise, Jordan and Powell [40] believe that a body concept develops from interacting with others. Anecdotal reports indicate that some children and adults with autism have an insecure body image
or totally lack body awareness. Russell [41] suggested that the ‘body schemas of persons with autism are poorly specified’ resulting in an atypical experience of agency. A sense of agency is a central aspect of human self-consciousness and refers to the experience of oneself as the agent of one’s actions. Based on his executive function account of impaired action monitoring Russell [42] put forward the hypothesis that individuals with autism are impaired in distinguishing between self and others. In contrast, a recent study by Williams and Happè [43] suggests that individuals with ASD are aware of their agency as indicated by their ability to monitor their own actions.

Support for a dominant role of the right hemisphere in the above processes is substantial. The right posterior parietal lobe is generally associated with spatial and bodily awareness [44,45]. Activation of right inferior parietal lobe correlates with a sense of ownership in action execution [46]. Additional research evidence [47] based on transcranial magnetic stimulation (TMS) supports the significance of the right temporo-parietal junction in the maintenance of a coherent sense of one’s body.

3.4. Theory of mind, emotions and self-awareness

An essential component of self-awareness is the ability to be aware of other minds. A multitude of studies have provided evidence that theory of mind is lacking or delayed in ASD, or develops differently than in neurotypicals [for a substantive review see 48]. Deficits in mind-reading may also affect the ability to reflect on one’s own mental states [49] resulting in diminished self-awareness. There is some evidence suggesting that the ability to think about one’s own thoughts depends on the same cognitive and neural processes as mindreading [50]. Equally, emotions play an important role in self-awareness. The development of the capacity to experience, communicate and regulate emotions is considered to be the most important event in infancy [51]. One of the main characteristics of autism is lack of empathy and emotional engagement with others [52,53]. Children with autism have difficulties with interpreting emotions, are deficient in processing their own emotional experiences and pay little attention to emotional stimuli in general [e.g. 54-57]. Due to this inability to empathize and emotionally engage with others individuals with ASD are totally focussed on their own interests and concerns.

A network of structures important for theory of mind processing includes the superior temporal sulcus and the adjacent temporo-parietal junction, the temporal poles and the medial prefrontal cortex [58; see also 59 for a review). Research evidence implicated the RH in theory of mind reasoning across various tasks [60, 61]. The neural substrates for emotions and empathy are complex [62] involving amygdala, ventral medial prefrontal cortex. Recent imaging studies point to an involvement of a ‘mirror neuron circuit’ for empathy [63,64].

3.5. Egocentrism/Allocentrism

In apparent contrast to the mentalizing impairment even among very high functioning individuals with ASD is their often-documented increased sense of self or total focus on the self [65,66,67] that is also reflected in numerous biographical accounts. The term ‘autism’ is de-
rived from the Greek word ‘autos’ (‘self’) and since Kanner’s time this focus on the self as atypical applies to all individuals with ASD. Extreme egocentricity was one of the diagnostic criteria for Asperger Syndrome proposed by Gillberg & Gillberg [68]. Frith and de Vignemont [69] suggest that there are differences to reading other minds depending on whether the other person can be understood using an ‘egocentric’ or an ‘allocentric’ standpoint. From an egocentric point of view other people are understood only relative to the self whereas from an allocentric stance the mental state of a person is independent from the self. These theorists suggest that individuals with ASD suffer from an imbalance between both point of views, ‘they are unable to connect an egocentric to an allocentric stance and can only adopt extreme forms of either’ [70]. This very detailed analysis of mindreading further illustrates the different and unique aspects of awareness of self and others in individuals with ASD.

3.6. Self awareness across time

Awareness that we are the same person across time, also defined as temporally extended self-awareness [71] is an essential part of one’s self-concept. Although the results of two recent studies [72,73] indicate that individuals with ASD have undiminished temporally extended self-awareness as assessed by the delayed self-recognition task, this task may not adequately measure self-awareness as suggested by Lind & Bowler [74]. Indirect evidence suggests that temporally extended self-awareness is impaired in ASD based on their problems with theory of mind as well as some aspects of temporal cognition [75]. Alternative explanations are the autobiographic memory difficulties [76] and also the well-documented language impairments in ASD. Language is a medium with which we monitor ourselves and it allows us to experience past, present and future [77].

3.7. Language and awareness of self

Conceptions about oneself and others develop from an early age and depend largely on the emergence of language. At around 18 months of age children start referring to themselves as ‘I’ and begin using the word ‘you’ for others, indicating a further development in their self-other awareness. According to Kircher and David [78] ‘the symbolic presentation of the self in language is the personal pronoun I’. Language difficulties such as pronoun reversal, use of third person perspective, impoverished inner speech, and impaired narrative have a negative effect on mental processes and also restrict certain aspects of self-awareness.

There is substantial clinical and research evidence of impaired pragmatic language use in children with ASD as indicated by pronoun reversal errors (‘I’/‘me’/‘you’) [79-82] reflecting general difficulties with their sense of self, as well as problems in self-other differentiation. Peeters et al. [83] suggested that the reason children with autism sometimes communicate from a third-person perspective instead of a first- vis-à-vis second person perspective is that in contrast to typically developing children they possess a non-social basis of self-other categorization. Use of a third person perspective also has consequences for attribution of mental states, and mentalizing ability in general. As argued by Northoff and Heinzel [84] a third person perspective is an indication of a fragmented image of self and other. Adults with
ASD also appear to have difficulties with first person pronoun usage [85]. Of particular interest might be the fact, that Hans Asperger often used to refer to himself from a third person perspective [86].

A fundamental role in self-awareness is attributed to inner speech [87] that is impaired in ASD [88]. When asked about the nature of their thoughts, a group of adults with Asperger syndrome reported mainly images and actions as their only inner experience and made no reference to inner speech or emotions [89]. Many individuals with ASD are visual thinkers and rely heavily on visualization to process information [90].

A recent fMRI study [91] provided evidence of underintegration of language and imaging in autism by showing that individuals with ASD are more reliant on visualization to support language comprehension. These authors suggest that cortical underconnectivity is the reason for the lack of synchronization between linguistic and imaginal processing in autism. Supporting these findings are the results of an imaging study on daydreaming [92] indicating that autistic individuals do not ‘daydream’ about themselves or other people. This study also points to a link between daydreaming and the construction of self and self-awareness.

In summary, language is of fundamental significance to self-awareness and necessary for forming a clear identity of self and others. Another important dimension in the formation of the self that is also dependent on language is autobiographical memory as well as the construction of a narrative self.

3.8. Autobiographical memory

Many influential theorists [93,94] associate the development of self with the emergence of autobiographical or episodic memory. The components necessary for a fully functioning autobiographical memory are a basic memory system, spoken or signed language, understanding and production of narrative, temporal understanding, self-awareness and theory of mind [95]. Autobiographical memory not only depends on these cognitive constructs but is also specifically concerned with events that have specific meaning to the individual. This personal significance evolves through emotions and motivations that are constructed in interaction with others. Autobiographical disturbances can arise from combined deficits in the realms of memory, emotion and self-related processing which are intricately connected, both behaviourally and neurologically [96].

The majority of components that make up an autobiographic memory system are impaired in autism. There is significant evidence that individuals with ASD have circumscribed episodic memory impairments, e.g. they have an impaired recall for personally experienced events [97-101]. As suggested by Millward et al [102] individuals with ASD remember real-life episodes less well than other people because they have no ‘experiencing self’. Wheeler et al [103] in their investigation of episodic memory in autism concluded that remembering of personal events requires the ‘highest form of consciousness, autonoetic consciousness (self-knowing), which is dependent of self-awareness’.
The prevailing view is that episodic memory is created in the neocortex and subsequently stored in the medial-temporal lobes and after a time becomes independent and is distributed in neocortical networks [104]. Whereas the left temporal lobe is dominant for the acquisition of new verbal information, episodic information involves mainly the right frontal lobes [105, 106]. Neuroimaging studies provide evidence for right frontal involvement in the processing of autobiographic memories [e.g. 107]. The RH is especially important for memories with emotional contents.

3.9. The narrative self

Many theorists [e.g. 108, 109] have highlighted the importance of the narrative self and argued that the autobiographical self is a similar construct as the narrative self. Individuals create their own identity by constructing autobiographical narratives or life stories [110]. The benefits of a personal or narrative self are significant; a narrative mediates self-understanding and creates coherence out of life's experiences. Narrative emerges early in development and narrative and self are inseparable [111]. The creation of a narrative self depends on various cognitive capacities, including working memory, self-awareness, episodic memory and reflective metacognition, a sense of agency, the ability to attribute action to oneself together with a capacity for temporal integration of events, a fully functioning pronoun system, an ability to differentiate between self and non-self as well as a sense of one's own body that is based on proprioceptive-motor processes [112].

The mechanisms responsible for each of the above dimensions are impaired in autism and as a consequence individuals with ASD have great difficulties in constructing a self-narrative. If autobiographical material cannot be provided, the narrative is disoriented and confused and in many cases is no narrative at all but only confabulation [113], which is often the case in autism. As a result, the narrative of individuals with ASD, and the self that is represented in this narrative, is quite vague and not representing the true self. Research evidence confirms deficits in narrative abilities in individuals with ASD [114-116].

Language and symbolic functions are localized in the left hemisphere, whereas narrative abilities are considered to be a function of the right hemisphere. There is a significant evidence for RH contribution to social language and many of the functions associated with autobiographic memory specifically those with emotional contents. In addition, narrative organisation depends on coordination of activity among various brain regions [117] and as suggested by Belmonte [118] malfunction in neural connectivity may be the underlying problem with autistic narrative.

To summarize, there is substantial evidence that the main components of self-awareness including self recognition, self-other differentiation, body awareness, theory of mind, inter-subjectivity, emotion processing, language (pronoun reversal, inner speech, third person perspective), autobiographical memory and narrative self are impaired in ASD. Our review of neural substrates underlying these processes has highlighted the significance of the Right Hemisphere.
4. Self neuropathology in ASD

From a neural point of view the self can be viewed as a complex and dynamic representation consisting of multiple brain networks [119, 120]. The origins of self begin in infancy and over the first several years of life normally developing children acquire an understanding of different dimensions of self and other. Deviant development in autism is likely to result in a cascade of developmental impairments including dysfunctional self-related processes as outlined above. Various brain regions have been indicated in the pathogenesis of autism including frontal lobes [e.g., 121] cerebellum [122], parietal lobes [123], hippocampus [124] and amygdala [125]. The extent of anatomical and functional abnormalities in autism points to a possible core dysfunction in neural processing. In addition, the vast amount of potential genetic risk factors suggests that multiple or all-emerging functional brain areas are affected during early development [126]. This theory is supported by widespread growth abnormalities in the brain of children with autism [127, 128].

In the following sections we will explore three neural theories implicated in the development of an atypical or different sense of self in individuals with ASD. Apart from the involvement of the RH in self-related processes a dysfunctional mirror neuron system as well as abnormal connectivity may have a role to play in the atypical developmental trajectories in ASD.

4.1. Right hemisphere hypothesis

The prefrontal cortex plays a vital role in the development of the self as it generates a sense of self and facilitates many links with other parts of the brain. Cognitive neuroscience studies have shown that the RH plays a special role in personal relatedness, which is intimately linked to the development of the self. Based largely on recent neuroimaging research evidence an increasing number of cognitive neuroscientists have emphasized the specific role of the RH in self-related functions [129-132]. Specifically the right dorsomedial prefrontal cortex seems to play a critical role in the development of models of the self [133]. This has been confirmed by several imaging studies, including a recent study of self-evaluation [134]. As described in the previous paragraphs there is substantial research evidence that the RH may be dominant for self-awareness and self-related functions. The psychiatrist and philosopher Iain McGilchrist [135] provided an extensive exploration of the dominance of the RH in self-related processes.

Elsewhere, Lyons and Fitzgerald [136] put forward the theory that RH impairment leads to a dysfunctional self-development in ASD. There is substantial research evidence linking Asperger syndrome to right hemisphere dysfunction [e.g. 137, 138]. The RH is dominant in the first years of human life when the major brain development during critical periods takes place. Results of a cerebral metabolism study in children (aged between 18 days to 12 years) showed that the RH is prominently activated, suggesting that the RH develops earlier than the LH [139]. The RH in implicated during early social interactions [140] including early attachment processes [141], maternal face and voice recognition [142] as well as the ability to view others in a similar way as the self [143]. The developing self depends on relations with
others and these early experiences are vital for the maturation of the right brain system. Substantial behavioural evidence of infants who later developed autism is supporting the theory of disrupted intersubjective behaviour. We argue that impairments in neurobiology affecting particularly the RH both cause and interact with defects in personal relatedness and later developing self processes.

4.2. Abnormal connectivity

The ‘Abnormal Neural Connectivity Theory’ proposes that autism is a distributed system-wide brain disorder that restricts the coordination and integration among various brain areas. The original positron emission tomography (PET) study by Horwitz et al. [144] found reduced correlations among frontal cortex, parietal and other brain regions and suggested that autism involves impairment in functional connectivity between frontal cortex and other brain systems. More recent studies proposed that autism is a disorder of neural underconnectivity [145], overconnectivity [146, 147] or both under and overconnectivity in which local connectivity may be relatively dense whereas long-range connectivity between brain regions may be reduced or abnormally patterned [146-151].

Studies of the cerebral cortex in autism show abnormalities of synaptic and columnar structure. Cortical minicolumns are fundamental units of cerebral cortical information processing. Examination of neurons revealed abnormalities in the size of cortical minicolumns particularly in the frontal and temporal lobes in ASD [152, 153] that could alter overall levels of connectivity within the brain. These findings are in accordance with the observed white matter abnormalities reported particularly in people with ASD [154]. A recent study using functional connectivity MRI (fcMRI) [155] provided further evidence of atypical enhanced functional connectivity suggesting that abnormal connectivity may be linked to developmental brain growth disturbances in autism.

These studies suggest that connectivity among diverse brain areas may be the core problem in autism. In autism the network connectivity through which various brain areas communicate with each other are limited, particularly the connections to the frontal cortex [156] which is dominant for self-related processing particularly in the RH. The network model of the self proposed by Stuss et al. [157] suggests that the self is hierarchically organized, with the highest level of the self involved in self-awareness being subserved by frontal lobes. Early developmental impairments in minicolumnar microcircuitry in the frontal cortex in autism could be the reason for the deficits found in higher order frontal processes [158] which are likely to result in fragmented self awareness and identity formation in autism.

4.3. Mirror neuron system

Another recent neural theory of autism suggests that a dysfunctional mirror neuron system may be fundamental to the aetiology of autism [159, 160]. The existence of mirror neurons in humans has been demonstrated by a number of EEG and imaging studies [e.g. 161]. Mirror neurons are activated in relation both to specific actions performed by self and matching actions performed by others, providing a potential bridge between minds [162] and might
have a role to play in self related processes. Mirror neurons may enable us to understand the actions of others by mapping the actions of other people to our own motor system and so allow a shared representation of actions. In addition to understanding the action of others this so-called ‘mirroring’ might also allow the automatic experience of the intention and emotion of the other person as suggested by Kaplan and Iacoboni [163].

Research has demonstrated that mirror neuron activity correlates with empathy [164] and social competence in general [165]. It has been suggested that mirror neurons are a prerequisite for the normal development of self-recognition, imitation, theory of mind, empathy, intersubjectivity and language [166, 167]. Furthermore, mirror neurons are likely to play a central role in self-awareness. To quote Ramachandran and Oberman [168] ‘they may enable humans to see themselves as others see them, which may be an essential ability for self-awareness and introspection’ (p.41). Developmental data suggest that there is higher imitative behaviour in children that can self-recognise, possibly facilitated by mirror neurons, in contrast to those who cannot [169]. Providing support for a RH hypothesis in self-related functions are recent imaging studies [170, 171] indicating that a frontoparietal ‘mirror’ network is associated with self-recognition processes.

Several recent functional brain-imaging studies have found evidence of mirror neuron dysfunction in individuals with ASD in social mirroring tasks [172], motor facilitation [173], and imitation [174]. A fMRI study [175] revealed that individuals with autism showed a different pattern of brain activity during cognitive tasks relating to self-referential processing. The authors concluded that a core deficit in autism might be related to the construction of a sense of self in its relation with others. Echoing Hobson [176] Iacoboni [177] suggests that primary intersubjectivity is the basis for the development of the neural systems associated with internal and external self-related processes. Failure or abnormal development of a fully functioning mirror neuron system in the autistic infant is likely to result in a cascade of developmental impairments including dysfunctional self-related processes.

5. Conclusion

The centrality of an impaired sense of self in autism has been the focus of research for many decades. The development of self-awareness is a complex process that involves integration of information from many sources and coordination across the brain systems involved in self-related concepts. A sense of self emerges from the activity of the brain in interaction with other selves. There is substantial evidence that early deficits in self-development including impaired relations with others result in a fragmented and atypical sense of self in ASD. In this review we have presented evidence that a great majority of self-related processes that are mediated to a significant extent by the right hemisphere are impaired in individuals with ASD. Additional lines of investigation indicate that an unintegrated sense of self in autism is also potentially associated with abnormal functional connectivity and an impaired mirror neuron system. Consequences of this atypical sense of self are the well-documented impairments individuals with ASD experience in the social and communication
domain. In contrast, there have been suggestions that this different sense of self might be a contributory factor to the significant talents and special skills present in a majority of individuals with ASD. Happè & Vital [178] put forward the notion that diminished self-awareness in ASD might be advantageous in the development of these special gifts.

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