

BODY DYSMORPHIC DISORDER IN PEOPLE WITH INTELLECTUAL DISABILITY: A BIO-PSYCHO-SOCIAL APPROACH

JARRETT BARNHILL, M.D.¹

¹University of North Carolina School of Medicine, Chapel Hill, NC

Body dysmorphic disorder (BDD) is classified as a somatoform disorder that combines body image disturbances with obsessive compulsive spectrum and somatic delusional disorders. This paper explores these dimensional features of each component of BDD among individuals across the spectrum of intellectual disabilities.

Keywords: body image, biopsychosocial model, delusional disorders, developmental disability, intellectual disability, mental retardation, psychiatric disorder

The core features of body dysmorphic disorder (BDD) include a distressing preoccupation with imagined physical defects, repeated but unsuccessful attempts to correct these defects, and symptoms of sufficient severity to interfere with social, psychological and occupational function.¹ In an earlier paper the author described two cases of suspected BDD in individuals with self-injurious behavior and mild intellectual disability (ID) in the first case and high functioning autism in the second. That review dealt with a subset of high frequency/low intensity self-injurious behavior that resembled BDD in normo-cognitive people. Unfortunately, a low index of suspicion for BDD and a tendency to focus clinical attention on comorbid anxiety and mood disorders makes it unlikely that BDD will be routinely diagnosed in people with developmental disabilities.⁸ One approach to the problem of under-diagnosis of BDD requires the use of a comparative methodology that compares and contrasts the core symptoms of BDD with other recognizable neuropsychiatric conditions. This paper addresses this boundary and outlines a bio-psycho-social approach to understanding syndromes like BDD in individuals with ID.

WHAT EXACTLY IS BDD?

BDD is classified as a Somatoform Disorder.¹ Clinically, BDD is characterized by a persistent preoccupation with imaginary defects in physical appearance and unwillingness to accept reassurances, contrary medical opinions, or contradictory photographic evidence. The most recent literature on the epidemiology suggests a prevalence rate of 1-3% of BDD in the general population.²³ Although surveys find an equal male-female gender distribution, clinical

populations tend to show significantly higher referral rates among females. This difference appears to result from culture-bound definitions of gender-specific attractiveness,⁵⁷ variations in presenting symptoms, comorbid psychiatric disorders,⁵⁹ and sources of referral, especially cosmetic surgeons and dermatologists. Because it shares many symptoms with other mental disorders, mental health professionals frequently overlook BDD and focus instead on more familiar disorders. As a result, BDD tends to be underdiagnosed.^{23,32,36}

Although reported during childhood, BDD is usually diagnosed during late adolescent and early adulthood.^{3,21} Unfortunately, the rise in post-pubertal prevalence rates creates several problems with differential diagnosis. Perhaps the most challenging requires the clinician to differentiate symptoms of BDD from issues related to sexual maturation, ongoing body image development and solidification of identity, and ever-changing adolescent social development. As a result, defining the boundary between gender-specific adolescent pre-occupations, the extent that physical appearance shapes psychosocial status and the core symptoms of BDD can present a real challenge.^{3,21} A second problem emerges because of the increasing incidence of most anxiety/mood disorders, eating disorders (ED), schizophrenia and obsessive-compulsive spectrum disorders after puberty. BDD is overshadowed in this age cohort as clinicians zero in on these more widely recognized conditions. Finally, puberty ushers in differences in gender distribution that skew clinical referrals towards females—higher rates of clinical referrals for both BDD and some comorbid internalizing disorders.^{8,38}

This simultaneous rise in the prevalence rates of both BDD and comorbid conditions compounds the tendency for BDD to get lost in the background. For example, the diagnosis of BDD is probably overlooked in males. This oversight results from a higher threshold for recognition, confusion with ED or personality disorders, or most probably, a lack of awareness that BDD exists in males.^{38,43} Under recognition of BDD in both sexes is not without risks, however. Studies suggest that untreated BDD contributes to long-term greater morbidity and plays a role in the increased the risk for suicidal behaviors.^{15,47}

Even though BDD lies within the category of somatoform disorders, there are substantial differences between BDD, and hypochondriasis and other somatoform disorders. In BDD, the focus is on an obsessive preoccupation with body image distortions rather than fear of illness, or exaggeration of sick role behaviors.^{2,47,54,56}

In contrast, individuals with hypochondriasis obsess about potentially devastating illnesses and seek medical reassurance, but generally do not engage in self-corrective efforts. Individuals with other somatoform disorders present with a wide array of physical symptoms that are often difficult to document or clarify. Individuals with either disorder display an exaggerated pattern of sick role and illness behavior. Although multiple diagnostic tests and medical procedures are common, these individuals appear to play out their problems with attachment issues with medical personnel through aberrant illness behaviors.^{2,54} Individuals with BDD on the other hand feel compelled to act on their own, and in spite of seeking reassurance from others, may hide the extent of their efforts to correct their physical defects, spending hours per day trying to correct or conceal their maladies.^{39,42}

BDD also appears to lie on a clinical continuum between obsessive-compulsive, eating, psychotic mood disorders, and delusional disorder, somatic type. Many experts believe that because of shared symptoms and high rates of comorbidity, BDD lies within the larger group of OC spectrum disorders.^{14,40} But there are major differences between these conditions. When compared to obsessive-compulsive disorder (OCD), BDD differs in focus (obsessive preoccupations with body defects) and patterns of compulsive behaviors (performance of various

grooming rituals or pursuit of corrective procedures). In addition, BDD differs from OCD in terms of the success of the specific compulsive rituals in reducing underlying anxiety or distress. In BDD, the grooming rituals appear to exacerbate rather than reduce underlying anxiety or intensity of beliefs.^{31,52}

There is a prominent lack of insight in most individuals with BDD. Lack of insight may result from the psychological defense mechanism of denial, but can figure prominently in certain neurological conditions—anosognosia (lack of awareness of a problem) and anosodiaphoria (lack of affective response to dysfunction) are examples.^{37,47} Insight also involves an attributional component—the belief that current symptoms are caused by an illness. Patients with BDD fail this test of insight by attributing their dysphoria and depression to their disfigurement, rather than their mental disorder. For example, someone may acknowledge anxiety or some compulsive symptoms but fail to link these symptoms to inappropriate concerns about physical appearance.^{14,31,40}

There is another variation of the theme. Insight is also useful in clinical settings to determine reality testing—if an individual can test or validate false beliefs and adapt erroneous beliefs to suite objective data or culturally accepted beliefs. For example, an assessment of insight is used frequently to evaluate psychosis in individuals with unusual beliefs or sensory experiences. In this context obsessions (with insight) differ from delusions where there is a failure to grasp the absurdity, oddness or alien nature of expressed beliefs. Insight in OCD compulsions also included an ability to see these beliefs and subsequent behaviors as abnormal, maladaptive, and ego dystonic (alien to personal beliefs). But things are not this simple. It turns out that insight is often compromised in patients with both BDD, ED, and nearly one half of individuals with OC spectrum disorders.^{12,29,42} For these individuals deficits in insight adversely affects their ability to appreciate or acknowledge either the pathological nature of body image distortions or the inappropriateness of their endless pursuit of corrective measures.^{31,39,40} Both problems may limit voluntary psychiatric treatment or if “persuaded” to enter treatment, compliance may suffer.^{42,54}

THE ROLE OF TEMPERAMENT AND FAMILY IN THE DEVELOPMENT OF BDD

Families represent both a microcosm of the larger culture and the initial training ground for acculturation. As the soil for development, families represent a core feature in our understanding of the development of both healthy and psychologically unhealthy development. The role of families in the development of mental disorders is incompletely understood. For BDD, family studies are emerging but to date, there is limited consensus on which developmental interactions are most relevant. It is apparent, though, that we need to abandon theories of linear causality (e.g., families cause schizophrenia) and divisive arguments of nature v. nurture, and focus instead on a transactional model of mutual and reciprocal interaction between family members and society. With these caveats in mind, it is intriguing to compare family dynamics across the spectrum of developmental and psychiatric disorders. The difference between "healthy" and "dysfunctional" families often centers on issues related to family structure, emotional attunement and support, conflict and problem solving skills, and flexibility to adapt to the changing needs of various members. These same forces also operate for parents and their children with developmental disabilities.^{3,28,46,47}

Temperament is a good starting point for this discussion. Families adapt to and modulate the temperamental traits of each member. When this process works, parenting styles match the child's temperamental style and families are able to assist in development. Problems, on the other hand, can arise from several types of mismatches. One mismatch grows out of an inability of parents to adapt or even acknowledge the child's temperamental style. Under these circumstances the child either adapts and represses or acts out, leaving little room for mastery.³⁵ Another scenario may involve children with extremely difficult temperamental styles who may overwhelm even the best parents. These difficult temperaments are often associated with neurological or other developmental disorders and can seriously tax the family's adaptive repertoire.

Behavioral inhibition is present in nearly 10% of children. These children tend to withdraw in novel settings, show lower rates of exploration and may be less likely to turn to non-family members for assistance. There is an expanding body of research that suggests behavioral inhibition is

related to other temperamental differences such as high harm-avoidance and low novelty seeking. Harm avoidance represents a temperamental continuum that includes behavioral inhibition, but also involves excessive cautiousness, as well as an exaggerated sensitivity to disapproval or potential negative consequences. Novelty seeking is a temperamental trait associated with increased motivational states, sensitivity to potential positive reinforcing properties, and high need for novel experiences. Permutations of these two traits also fit into internalizing-externalizing and "neuroticism" dimensions. From this viewpoint BDD generally reflects high levels of harm avoidance, low levels of novelty seeking, high levels of neuroticism and predominantly internalizing symptoms.^{35,48}

Behavioral inhibition seems to be a core temperamental feature of several mood and anxiety disorders. This common neurobehavioral substrate underlies social anxiety, panic and other anxiety, mood, OCD^{14,40} and ED.¹² How these temperamental traits develop into different psychiatric disorders (BDD v. mood disorder versus social anxiety disorder) is not well understood. There is little doubt that the evolution of primary psychiatric disorders is the result a complex interaction between biology and psychosocial ecology. For example, behavioral inhibition and emotional reactivity contribute to a child's vulnerability to dysfunctional family systems. Difficulties with novel demands or experiences, shyness, sensitivity to disapproval, and the perception that the outside world is threatening close down many options for corrective interactions.⁴¹

It is safe to assume that the developmental pathway towards BDD differs to some fashion from the one that leads to social anxiety or ED. In addition to family and temperamental influences, factors contributing to this evolution involve genetic or neurobiological differences such as the patterns of information processing and brain activation (i.e., such as specific aspects of body image or affective reactions to facial appearance). These mutual interactions influence, intensify or reinforce these neurobiological temperamental differences.^{8,14,36}

From this point of view, vulnerable children growing up in families who define psychological well-being in terms of physical appearance or excessive conformity to existing cultural values of attractiveness are at particular risk.^{15,35} Another

piece of the risk puzzle may come from children (including those with ID) who live in families that also overemphasize attractiveness, and are exquisitely sensitive or overreact to specific physical features- imperfections that are defined as “ugly” or “defective.” Children with temperamental traits such high harm avoidance, slow adaptability or behavioral inhibition, are more apt to become enmeshed or “buy into” this world-view. In addition, their sensitivities to negative or aversive consequences (harm avoidance), shyness or reluctance to rely on non-family members for social support (avoidance behaviors) may close a range of developmental options.^{8,28,48}

CULTURAL FACTORS

Cultural factors also play a role in the development of BDD. From an aesthetic perspective, the core symptoms of BDD are shaped by culture-bound concepts and gender stereotypes of attractiveness—hence the apparent overlap with ED.¹² Since each member of a society is immersed in culturally-defined ideals of attractiveness, it is natural to assume that these cultural influences shape the dynamics of child rearing and family development.¹⁰ But even though the development of individuals is shaped by these attitudes, there is still considerable variance in the level of acculturation into these belief systems. As noted, the vulnerability to BDD is the result of a transactional process that also incorporates individual or temperamental differences. For example, individuals with high levels of social anxiety, harm avoidance or low novelty seeking and avoidance behaviors might express greater vulnerability to social anxiety and greater attachment to the prevailing belief system—less risk taking or overt challenges. As a result, these individuals might be more attuned to and willing to accept social cues of disapproval or more likely misinterpret or misattribute problems in interpersonal relationship to their “ugliness” or deformity without seeking alternative sources of validation.^{14,15}

Considering the broad range of attitudes and beliefs about beauty and attractiveness, there remain significant concerns about the reliability and validity of BDD across cultures. To date there are no studies comparing the clinical features of BDD in industrialized and traditional or non-industrialized societies. It is also risky to extrapolate differences in the prevalence rates of

body image disorders (ED v. BDD) across cultures. For example, cross-cultural studies of OCD suggest a common theme (repetitive behaviors) but differences in symptom focus.^{2,15,47} Where BDD fits into this continuum is largely unknown.

PSYCHOBIOLOGY OF BODY IMAGE, BDD AND ED

BDD represents a fundamental disturbance in body image (imagined disfigurement) and a compulsion to correct these defects regardless of consequences. The relentless attempts at self-correction by an affected individual may eventually result in self-mutilation or disfigurement.^{31,41} Unfortunately, reassurance has little impact and many individuals turn to medical or surgical solutions. Dermatologists and cosmetic surgeons are frequently consulted to correct facial or skin-related defects. For most, surgical procedures are ineffective and tend to exacerbate the patient's anxiety, and may result in legal action for failed procedures or additional damage. Neither solution is effective for reducing the obsessive preoccupations or ritualistic grooming.^{39,42}

ED (anorexia and various forms of bulimia) present with a combination of body image disturbance and syndrome-specific eating/feeding behavior. Using either of these features as a starting point, clinician can subtype ED based on the nature and intensity of compulsive/ritualistic dieting (restricting), obsessions with calorie computation, distorted views of the relationship between calories and weight gain, bingeing/purging behaviors, and the role of exercise regimens (calorie computation).^{12,48,54} Cognitively most individuals with ED lack insight into the pathological nature of their disorder; resist reassurance or evidence contrary to their belief system; misattribute problems with interpersonal and psychosocial difficulties to their distorted body image; or distort the effects of even moderate caloric intake on weight maintenance.⁴¹ Individuals with anorexia nervosa-bulimia nervosa are relentless in their need to correct their body image distortions. This drive to correct a distorted body image overrides most health concerns and social, educational, or occupational needs of the individual—pre-occupation and relentless compulsive need to correct self-perceived appearance eventually results in progressive social isolation and alienation. Severe health

complications and death are surprisingly common outcomes.^{12,15,41}

Within the broad category of ED, there are temperamental differences in the degree of harm avoidance, impulse control, affect regulation, and psychiatric comorbidities. In this respect the spectrum of ED, especially bulimia with bingeing-purging bears a phenomenological relationship to other compulsive spectrum disorders, borderline personality disorder, and addiction behaviors.³³ Restricting forms of anorexia falls at the other end of the continuum—higher level of harm avoidance, less impulsive, and a different pattern of psychiatric comorbidity.^{12,41}

Individuals with BDD often present with a similar pattern of preoccupations but differ in terms of their areas of focus—a general shift from the body as a whole to specific regions. There appear to be significant gender related differences as well. For example, females with BDD shift their focus away from fat distributions and preoccupations with being overweight and focus on anatomical limited areas (face, hair, etc).^{38,46} On the other hand, males with BDD present a more confusing picture in that they are more likely to share an expanded anatomical focus with other ED.^{38,43} In addition to obsessive preoccupation with body image distortions, males with BDD may feel compelled to alter or spend hours per day trying to correct or conceal their maladies. Their relentless attempts to correct eventually result in serious health consequences—self-mutilation, anabolic steroid abuse or wear and tear injuries.^{38,45}

Finally, reassurance has little impact on their anxiety and obsessive pre-occupations. Many individuals eventually turn to medical or surgical solutions. Dermatologists and cosmetic surgeons are frequently consulted to correct facial or skin-related defects, breast size, and abdominal anatomy. For most, surgical procedures are generally ineffective and in many situations, may exacerbate the patient's anxiety, and may resort to legal action due to either a failed procedures or additional damage.^{14,42}

PSYCHOBIOLOGY OF BDD, OCD AND DELUSIONAL DISORDERS

Persistent and intense obsessive convictions of and preoccupations with perceived body defects, compulsive attempts to correct these misperceptions, and sensitivity to SSRIs and

cognitive behavioral therapies characterize both BDD and OCD. Although OCD is frequently associated with other anxiety and mood disorders, recent studies suggest that some obsessions in BDD also lie on the boundary with delusions. The certainty of beliefs or fixed ideas, lack of insight into the pathological nature of these symptoms, and general disregard for contradictory evidence define this boundary. The presence of OC symptoms in individuals with schizophrenia or psychotic affective illnesses also supports a dimensional rather than categorical construct.^{31,50}

Individuals with BDD tend to misinterpret ostracism by friends or professionals as confirmatory evidence or devaluation by others. Interestingly, the individual denies, underestimates or minimizes the psychopathological nature of their symptoms. At this point, the patient's preoccupation with imagined disfigurement takes center stage and is considered to be the sole cause of all social distress. The compulsive need to correct these misperceived defects becomes the primary life focus. Unfortunately, reassurance or contradictory evidence have little impact. Ultimately, the patient with BDD feels increasingly misunderstood, maligned and alienated by exasperated family members, friends or even medical providers. The patient misinterprets avoidance as rejection due to their disfigurement. For many, the intensity of these convictions (fixed or overvalued ideas) can reach the boundaries for delusions—absolute certainty of dysmorphology, refusal to accept contradictory evidence, and sense that others are “talking about their defects.”^{37,46}

Fifty percent of individuals with BDD lack insight into their misperceived body image disturbance. Most do not respond to reassurance or dissuasion. Many patients with severe BDD also persist in their conviction that everyone can readily see and reacts adversely to their disfigurement. The intensity of these symptoms contributes to serious impairment in psycho-social functioning and chronic interpersonal difficulties—lower rates of marriage and occupational status. Isolation, alienation, and intensification of symptoms may eventually result from increased preoccupation with and frustration over a failure to correct the defect.^{31,37}

In spite of the intensity of these fixed ideas, there is little evidence of a formal thought disorder or psychotic mood disorder. For example, individuals with BDD lack many of the core

features of most schizophreniform disorders—thought disorder, affective blunting, complicated hallucinations, and associated neurocognitive deficits. But the differential diagnosis of psychotic mood disorders can be more complicated. At times it may be difficult to distinguish “delusional” BDD from delusional body image or social distortions associated with melancholic/psychotic depression, including premorbid histories of social anxiety, or OCD. Although certainly not an absolute distinction, the severity of depression may provide clues. For example, the presence of delusions is more directly correlated with the severity of the mood disorders (mania or melancholia). But a clinician unfamiliar with BDD may feel these delusional beliefs are part and parcel of a psychotic mood disorder and proceed to treat accordingly. But there are key differences in age of onset, clinical course, neuroendocrine, and treatment sensitivity.⁴⁶ The presence of body image disturbance in milder forms of depression might suggest comorbid BDD. In either scenario, there is a considerable level of functional impairment (chronic and often treatment refractory illness) as well as increased suicidal risk.^{31,39,42}

NEUROSCIENCES AND BDD

Even though both BDD and anorexia nervosa/bulimia nervosa involve disorders of body image, there are phenomenological differences between the two. For example, a key distinction between BDD and ED is the topographical differences in symptom focus. Individuals with BDD tend to selectively focus on the face, hair, nose, or other facial characteristics. Individuals with anorexia nervosa react to their entire body gestalt—either the distribution of fat or gross distortion of body shape. Recent neuroimaging studies suggest that ED are associated with significant problems in hypothalamic and parietal-occipital networks. In contrast to these findings, neuroimaging studies of individuals suggest that individuals with BDD focus more on facial perception than a global body gestalt, including affective valence assigned to facial image, and facial attractiveness.^{8,14,46} More specifically, the pattern of aberrant activation points to involvement of the inferior aspects of temporal and parieto-occipital regions that may differ from the more global processing of body image found in ED. The tendency to zero in and remain focused on facial defects (over-selective attention) also suggests that in addition to

aberrant activity in posterior regions (temporal and parieto-occipital structures) linked to body image, BDD might also represent disturbances in attention networks, and stimulus over-selectivity involving facial processing.^{11,13,19,44,46,58}

The perceptual processing of facial features and body image involves a complex circuit that includes both cortical and subcortical pathways. The fusiform gyrus (inferior temporal cortex) is the critical brain region for processing faces. This region is also linked to mesial temporal structures that are sensitive to emotional signals (nonverbal facial expression). This region is also involved in assigning affective valence to these highly processed images. Global body image on the other hand, tends to involve posterior regions (the confluences of posterior temporal parieto-occipital regions). These regions are involved in spatial/constructional tasks, attention, and body schema. Both “regionalized” processing modules share and ultimately integrate this information with other limbic and prefrontal regions.^{19,26,53} This circuit links perception of self and others with memory, emotional responses, past experiences, and other context related cues. Destructive lesions in this region can result in a failure to recognize specific faces and attach a name to the facial representation but not affect one’s ability to recognize that the visual cue is a face, or prevent the individual from using other cues (dress, voice, or nonfacial cues) to make an accurate identification.³⁴ Similar disorders can affect one’s capacity to recognize their image (autotopagnosia).^{26,44,55}

Individuals with BDD or ED on the other hand, have no trouble recognizing themselves or others but are convinced that something is wrong. They also link this faulty perception to a disgust reaction in response to their own appearance. The decidedly negative emotional reaction or valence (negative or disgust reactions) tends to be similar—both site-selective (BDD) and global misperceptions (AN) trigger a negative affective reaction and a sense of disgust.^{2,19,22,47,49} The origins of this shared disgust reaction suggest that both disorders share a similar connection to lateralized limbic or medial temporal activation that process affective meaning of highly processed sensory stimuli. The amygdala is a crucial component of this processing and affective reaction system.⁵

Neuropsychological studies of individual with BDD also suggest a bias in processing ambiguous

stimuli with exaggerated negative affective responses. The intensity of the negative affects attached to the visual representation of body image apparently overrides realistic self-appraisal (reality testing) or input from significant others. Under normal circumstances, the prefrontal cortex is involved in self-appraisal and mediating internal affective state with external contingencies (social validation or reality testing). The combination of faulty self-perception and a perseverative, negative affective response states attached to body image suggest a disturbance in multiple brain circuitry (coherence).^{44,49,53}

The process becomes more complicated when we add behavioral inflexibility to the mix. Inflexibility and compulsive-repetitive behaviors are central to OCD, ED and BDD. The introduction of compulsive behavior to the perceptual problems seen in BDD and ED shifts the focus of neurophysiology towards the fronto-striatal pathways. Functional neuro-imaging studies of individuals with ED suggest different patterns of activation in posterior brain regions.^{13,14,19} Functional imaging studies of BDD also suggest posterior and mesial temporal involvement. OCD and compulsive/perseverative behaviors occur in conjunction with fronto-striatal abnormalities.^{13,14,46} Yet, in spite of these regional differences between specific syndromes, there is substantial common ground and overlapping neurocircuitry. A similar overlap in neurocircuitry is also found in many of the comorbid conditions associated with OCD, BDD and ED—mood, social anxiety and other anxiety disorders.^{12,41,46}

But these neuro-imaging studies also support differences between OCD, BDD and ED. These differences appear most prominent through impairments in orbito-frontal, cingulate, and caudate pathways in OCD. These dysfunctional networks appear less involved in BDD/ED (body image disturbances) where posterior changes in regional brain activity are most prominent.^{13,19} Mood and other anxiety disorders involve overlapping pathways but may have prefrontal and mesial temporal problems.^{5,6} Interestingly, the data on Somatoform disorders is less extensive.^{2,54}

These regional variations in functional and anatomical neuro-imaging studies between specific syndromes raise intriguing questions about the boundaries between comorbid conditions. These differences may partially explain why comorbidity is a marker for greater

severity or treatment resistance of the primary psychiatric disorder. Perhaps, comorbid conditions represent a more generalized level of functional impairment. On the other hand, different patterns of brain activation may provide insights into an age old problem—why do so many disorders share similar family and social dynamics.

From a neuropsychiatric perspective, BDD appears to involve brain regions linked to compulsive or other ritualistic behaviors and disturbances in self-perception. Lack of insight is another confounding feature. Many individuals with BDD, ED and some obsessive-compulsive spectrum disorders seem to be either unaware or not deeply troubled by their disorder. Rather they seem convinced that something is amiss, but often fail to connect these misperceptions to a mental disorder. This pattern of cognitions and resulting minimization of the pathological nature of their symptoms is a common feature of delusional disorders. In addition to individuals with delusional disorders, those with BDD and anorexia nervosa also share fixed-ideas, over-valued convictions, and a diminished capacity to use consensual validation or contrary evidence to alter beliefs or perceptions.^{14,20,31,53}

Neurobiologically, these deficits suggest disruptions in the interaction between executive or fronto-striate networks and perhaps to a lesser extent, posterior networks involved in complex sensory perception. The prefrontal cortico-striate pathways also play a role in the regulation of limbic connections that are involved in affective responses (negative or disgust reactions) to both internal and external input.^{11,22,37} There are also differences in terms of brain lateralization. Most delusional disorders involve subcortical and nondominant hemispheric dysfunction (usually the right). From neurophysiological studies, it is apparent that over-activation of the greater amygdala (nondominant) is associated with fear, avoidance, rage, and increased sensitivity to disgust or other negative facial expressions. This activation pattern also plays a role on behavioral inhibition, panic, and post-traumatic anxiety. Lack of insight or awareness of illness, and limited capacity to use an ongoing stream of information to change behavior suggest a different neurocircuitry.^{16,22,26,46}

BDD and other body-image disorders involve not only disturbances in self-perception but also a tendency to misinterpret a range of social cues.

There is growing evidence that such complex perceptions involve the interpretation of facial cues (in BDD) and body image (in anorexia nervosa), while social cues have more in common with delusions.^{50,51,53} Recent studies suggest that nearly 50% of individuals with BDD have an illness that reaches delusional proportions and express a more severe form of the disorder. The association between increasingly severe illness and the presence of delusions is observed in individuals with somatic delusions in psychotic mood disorders and chronic schizophrenia. As noted earlier, there are significant boundary issues between some forms of obsessions or fixed ideas, and delusional beliefs—including overlapping areas of cognitive dysfunction.^{16,17,41,51} Even though somatic delusions can focus on defective body image, there are other dimensions that may be helpful in the differential diagnosis—disturbances in effectiveness (grandiosity-worthlessness-hopelessness continuum), physical appearance (beauty-decay spectrum), perception by others (grandiose-deprecatory referential thoughts), and the presence of other delusions or hallucinations.^{9,20,30,46,50}

Individuals with severe BDD display symptoms that also resemble content specific delusions (such as Capgras or delusion of imposters). Although reported in schizophrenia, content specific delusions are more commonly reported in patients with Alzheimer's disease and lesions in the nondominant fronto-parietal region.^{18,30} Neurological disorders that affect the temporal lobes may also produce changes in body image. Autotopagnosia is an example.¹⁶ Even though most patients with BDD do not have specific neurological lesions to account for their symptoms, there are similarities in affected brain regions. Perhaps we can learn more about the neurobiology of BDD by investigating content specific delusions, especially those individuals who lack symptoms of severe dementia, schizophrenia or other psychoses.

BDD AS A DEVELOPMENTAL DISORDER

We currently diagnose BDD based on a collection of descriptive criteria. The clinical reality is that BDD lies on a relative continuum (dimension) with disorders of body image, OC spectrum, and probably delusional disorders. BDD is also a disorder in which comorbidity is probably the rule rather than the exception. Differentiating BDD from these comorbid

conditions may require considerable clinical skill and patient cooperation/ self-reflection to elicit specific symptoms (criteria).⁸

Problems remain even as we step beyond a listing of criteria and look at neurodevelopmental or other risk factors. The neurobiology of BDD suggests an interaction between genetic factors, temperamental risk factors and psychosocial environment during normal development. Precisely how social and cultural factors shape neurobiological risk factors such as behavioral inhibition into a specific syndrome remains unknown.^{5,6,8} The relationship between family dynamics and temperament illustrates this point. In contrast to the developmental dance of most families, those associated with BDD may lack mutual nurturance and capacity to match the temperamental characteristics of their growing children. When coupled with behavioral inhibition, this dynamic may bias development in the direction of internalizing disorders. The final step towards BDD is still unclear but is probably related to different underlying brain substrates—the way the human brain visually processes facial features or emotional reactions to these complex social stimuli.^{14,27}

Our understanding of the complex interaction between the neurobiology of behavioral inhibition in individuals with intellectual and adaptive deficits is in its infancy. If we take a larger view, however, it is apparent that culture and social values play a key role in shaping not only attitudes towards ID but also a sense of hope or fatalism about psychosocial outcome.²⁵ For example, families are the principle support system for many individuals with ID.^{5,6} The families of children with ID face a series of challenges throughout their life span. Depending on the level of ID or associated neurological disorders, these challenges may last well beyond those faced by families of "healthy" normo-cognitive children. Even in the face of these difficulties, successful families develop a range of healthy, growth promoting strategies and are sufficiently resilient to find workable resolutions to most developmental challenges. For other families, a child with developmental disabilities has a devastating impact and efforts to "correct" or "undo" the psychological grieving for the lost perfect child can unleash a wave of compulsive tests and/or interventions. In spite of these differences in family dynamics, our understanding about the ways each of these psychological forces

interact with social values and cultural expectations about ID affect the development of BDD in children is still largely unexplored.²⁷

The recognition of mental disorders is not a straight forward process for individuals with ID. Cognitive and communication deficits have a significant effect on the capacity of an individual to communicate internal states, perceptions or adapt to stress. At the same time, the physiological state associated with most mental disorders influences and is influenced by these same problems. The convergence of behavioral inhibition and impairments in adaptive skills and other executive functions increase an individual's vulnerability to stress by limiting the individual's capacity to generate alternative solutions. Depending on the level of ID, this same cluster of deficits can also prolong reliance on significant others and affects the quality of attachments and level of autonomy.^{4,6} In addition, the combination of cognitive deficits and behavioral inhibition puts additional pressure on support systems. For children already vulnerable to internalizing disorders, the process of developmental weakening of these dependent relationships may generate significant stress. The stress of late adolescent separation and striving for independence is perceived as potential disruption of these attachments. This same convergence of factors also adds to the risk for emerging mental disorders, including BDD.^{4,25}

The question arises—how do we recognize high risk children in high risk families? Many clinicians work with children who in addition to ID and developmental disabilities have significant orofacial or dysmorphic features. Many of these families are quite assertive in pursuing and obtaining cosmetic or reconstructive surgery. So, is this a family problem or are the children willingly compliant or perhaps encourage parental obsessive behaviors? The answer requires a systematic look at specific family dynamics rather than generalizing or “pathologizing.” Firstly, what are the underlying motivations for interventions such as corrective surgery? It may turn out that families at risk for children with BDD differ from those who strongly advocate for corrective or cosmetic surgery for cleft palates or even Trisomy 21. At risk families may display an unwillingness to accept the advice of close contacts (teachers, etc.) or physicians who contradict the family's assessment of defectiveness.

What happens if the procedure fails to satisfy the family? At risk families may not be satisfied regardless of objective outcome measures. Lacking satisfactory results the family may obtain serial assessments and procedures. Their persistent willingness or compulsion to subject the child to repeated surgical or dermatological procedures is a potential red flag. Their relentless pursuit of intrusive or often painful medical-surgical treatments may represent a “BDD by proxy”—perhaps similar to “Munchhausen by Proxy.”^{3,4} Under these circumstances, attachment and psychological support are wedded to the child's capacity to play the role of a dutiful but deformed patient. These traits may be warning signals that might help clinicians differentiate high-risk families from those grappling with serious orofacial defects.

Why do families with similar dynamics and children with at risk temperaments present with different syndromes—internalizing disorders such as eating, obsessive-compulsive spectrum disorders, social anxiety, and other somatoform disorders? From a cultural perspective, family and culture define the global context for BDD and exert a significant influence on vulnerability. The evolution towards a specific syndrome differs due to the interactive effects between these global family issues and a specific genetic or neurophysiological substrate. Even though we have spent much of this paper defining phenomenological boundaries between these overlapping syndromes, the factors that eventually develop into emergence of BDD rather than anorexia nervosa are probably “idiosyncratic.”²⁷

This problem is compounded for people with ID. It may be far more difficult to define boundaries that segregate BDD from ED, OCD, and psychotic disorders across the spectrum of IDs.^{5,8,25} Similar problems arise when the symptoms of BDD occur in individuals with known neuropsychiatric disorders. The boundary between BDD and delusional disorders illustrates this problem. It is well established that delusional beliefs are linked to cognitive abilities and stage of development. Delusions in many individuals with ID are often simple, not well elaborated or embellished, and can be confused with fixed ideas or obsessions.²⁵ The presence or absence of insight is also unhelpful in the differential diagnosis.^{14,25,46} For example, nearly 50% of normo-cognitive patients with BDD present with

delusional forms of the disorder. These individuals need to be differentiated from people with somatic delusions in schizophrenia or psychotic mood disorders.^{18,24,40,41} This process is far from straightforward in individuals with moderate to severe ID. Similar problems arise in the differential diagnosis of organic disorders of body image, social perception, and content specific delusions where the impact of ID can be profound—significant impairments in the individual’s ability to test the validity of these perceptions, or make corrections in patterns of social misattributions.^{8,25}

Deficits in executive functions (attributed to functional deficits in frontal lobes) are part of the adaptive deficits reported in individuals with ID and/or autism. As such, defining the boundary between fixed belief, delusion, or perceptual distortions in these individuals may not be a straight-forward task. There are several subcortical structures involved, but only one nondominant hemisphere associated with many “organic” or content specific delusions.^{20,30,51} This model may be helpful in defining the nature of delusions in people with ID. For BDD, the problem expands to include problems with an affectively charged negative self-image (body image) and a compulsive need to correct this defect. The idea that some forms of self-injurious behaviors may be the most likely presentation for BDD is consistent with the neurobiological construct of delusions, body image disturbances, lack of insight and other compulsive-ritualistic behaviors.^{7,8,14} Resolving these issues will require a detailed analysis of target behaviors, search for comorbid mental disorder, and the eventual use of more sophisticated functional neuro-imaging for individuals with suspected BDD who present with self-injury.

CONCLUSION

In closing, no single model captures the complexity of most psychiatric disorders in individuals with ID. It is apparent that we need to explore syndromes such as BDD in a fashion that combines neurobiology with temperament, individual development, family systems and socio-cultural factors. But such a model poses a real problem for busy clinicians. Even though my intent was to avoid the pitfalls of linear causality and to look at BDD from many angles, the amalgamation of many models may create an intellectual version of “Frankenstein’s monster.”

I am still uncertain if we can consistently and reliably recognize BDD in people with ID unless we expend considerable energy in the differential diagnosis. We can start with global symptoms such as self-injury or body image disturbance and then proceed down a decision tree to define the boundaries between BDD and other psychiatric disorders. It may turn out that BDD represents a metaphor for the problems all of us face when try to understand people with dual diagnoses. Yet in spite of a certain fatalism that accompanies such an exploration, this paper reiterates the essential need for a systematic and multidimensional approach to psychiatric diagnosis.

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CORRESPONDENCE: Jarrett Barnhill, M.D., Director, Developmental Neuropharmacology Clinic, University of North Carolina School of Medicine, Chapel Hill, NC 27599-7160; email: Jarrett_Barnhill@med.unc.edu