

# Dysmorphophobia in a Diastrophic Dwarf: A Psychiatry-Dermatology Liaison Approach

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## INITIAL COMMENT

This is a case study of a 30-year-old male diastrophic dwarf who complained of hypertrichosis on his back and nose of 3 years duration. He had no previous psychiatric history, and although the distortion he described on his back was not apparent to others, there was a minimal amount of dark hair growth on his nose. He had unresolved conflicts concerning his disproportionate arms and legs and had sought surgical correction for many years from many orthopedic surgeons to no avail. After a diagnosis of dysmorphophobia was made by a psychiatric consultant, weekly visits with the dermatologist were arranged for supportive psychotherapy combined with electro-epilation of the hair growth on his nose. The patient experienced a steady improvement and discontinued treatment after a few months with the understanding that he could return as needed.

The patient presented to the dermatology clinic while the author was an intern rotating in dermatology. The author worked with both the dermatology and psychiatry attendings in evaluating this unusual case and then presented it during a dermatology grand rounds. This case serves to illustrate the interesting interface between psychiatry and dermatology which is a growing area of clinical psychiatry-dermatology liaison work (1,2).

## INTRODUCTION

Dysmorphophobia, otherwise known as body dysmorphic disorder in *DSM-III-R* (300.70), the dysmorphic syndrome (3), or dermatologic nondisease (4) is an uncommon psychiatric disorder which refers to a distortion of the psychological body image. It was originally described in 1886 by Moselli as "a subjective feeling or physical defect which the patient feels is noticeable to others, although his appearance is within normal limits" (5). It is presently defined in the *DSM-III-R* as a preoccupation, not of delusional intensity, with some imagined defect in appearance. If a slight physical anomaly is present, the person's concern is grossly excessive.

Dwarfism by contrast is an undeniably real distortion of the body. As reviewed by Brust, many authors have seen the potential for emotional difficulties in dwarfs. Their small size attracts unusual treatment by relatives (6,7), teachers (8), and peers (8,9). For example overprotection (6,8,10), being treated

as if they were younger (7,8,9,11) and being deprived of heterosexual experiences (11,12) are common. Various cultures today view dwarfs as reincarnations of the devil, or associate the birth of a dwarfed child as punishment from God or the result of an eclipse or evil eye (13). Often they are shunned, abused or placed into orphanages in different cultures (13). Frequently personality traits include immaturity and dependency (12,14), extroversion (15), latent depression (11,14,15), and the absence of overt aggressive behavior (11). Ego defense mechanisms include denial (9,12), exaggeration (11,14), obsessive compulsive defenses (11), withdrawal or social isolation (9,11). Other coping mechanisms include the role of mascot (9,12,16), humor (12) and scholastic achievements (17).

The labels "midget" and "dwarf" differentiate two main types of little persons. Midgets have proportionate short stature while dwarfs are disproportionately short statured. Diastrophic dwarfism is a form of short limbed dwarfism in which the head and trunk are grossly normally sized. It was first described by Lamy and Maroteaux in 1960 (18). The term "diastrophism" is a Greek derived geographic term meaning twisted, tortuous or crooked and refers to the process by which the earth's crust is distorted to form continents, basins, mountain ranges, and so forth. Lamy and Maroteaux apparently selected this term because it describes the twisting deformities of the feet, hands, back and ears which characterizes this autosomal recessive disease. It is an intrinsic bone dysplasia where the primary defect is thought to be an error in chondrogenesis involving collagen formation. The prognosis is usually good for those surviving infancy. Severe kyphosis is a common problem causing impaired pulmonary function and frequent respiratory infection. Limited mobility is another characteristic because of the subluxed and dislocated hips and knees from ligament laxity (19).

There has been no random sample studies of the psychological status of dwarfs to this date. One of the largest and most recent studies by Brust, et al. (20) suggests that there does exist a sizeable group of dwarfs who function well as psychologically healthy and productive members of the community. Another study by Folstein, et al. (21) showed 30% of the dwarfs studied had an abnormal level of psychiatric symptoms that correlated with unemployment and poor education. Documentation of an attempted suicide in a depressed adult male diastrophic dwarf was made by Walker, et al. (19) and at least three successful suicides by adult achondroplastic dwarfs have been reported in the past few years (13). No clear estimate of the amount of psychiatric illness in dwarfs can be made at this time as the withdrawn and isolated individuals who do not belong to Little People of America are for the most part not accessible for study.

#### REPORT OF A CASE

A 30-year-old short limbed male dwarf was seen at the dermatology clinic complaining of hypertrichosis on his nose and back. He had noticed the hair on

the tip of his nose for 1 year and had been shaving it once per month. He did not feel it was "natural" and described an incident in a restaurant where his male acquaintance suddenly broke out in laughter exclaiming, "I just noticed you have hairs on your nose". He had been aware of the hair on his back which he described as "beastly, excessive, uneven and disproportionate" for the past three years. He considered the hair on his back to be disproportionately covering his shoulder blades. He stated, "anything that is not evenly distributed is odd." He described his experiences exercising in a special gym for handicapped people where after he began perspiring he removed his T-shirt and soon afterwards he felt the other people around were moving away and trying to avoid him. Since then, he had never taken his shirt off in public and had exercised at the gym less frequently. When asked if he had compared his body hair to other men's he conceded that while some men did have as much hair as he did, their's were evenly distributed. He felt that if all the hair on his back and nose were removed, he would look more normal and his difficulty in attracting women would improve.

He had been wheel chair bound for 4 years because his legs became weak and doctors told him he had no more cartilage left. He began having difficulty with mobility at age 17 and pointed out that with his 14 inch legs it was difficult for him to support his weight. During the past 13 years, he had seen numerous orthopedic surgeons in an effort to obtain extended limb lengthening of his arms and legs to no avail. Over the past few years he began thinking that it would be wonderful to have surgical procedures on both his arms and legs to get rid of those "ugly toes and fingers" and have artificial substitutes attached. He began thinking of himself as an amputee with stumps for both legs and arms and surgery offered the best chance of improvement. During the preceding 6 months, he had been fitted with a prosthesis that was designed to accommodate his feet without any surgical change and that added another foot to his height. The patient's understanding was that if he demonstrated to his surgeon that he could walk using the prostheses then the amputations would be performed. He was not satisfied with the prostheses alone because he didn't want to have to be "pretentious." By that he meant he did not want to have to pretend that he was an amputee. He said that he would feel humiliated if anyone should discover what was going on and, therefore, he wanted surgery so that he could feel justified in wearing the prostheses. He made the analogy that if he were bald, he would not want to wear a toupee but rather he would grow bald gracefully. He felt that by having the surgery was a sort of graceful way of adapting to the prostheses. One month prior to his presentation after he demonstrated that he could support himself and walk using the prostheses and a cane with a fair amount of difficulty, he was told by his surgeon that such amputations were a poor medical and orthopedic risk in his case. He then ended his relationship with his surgeon bitterly. He stated, "I felt like I was their learning tool. They just lied to me. They wanted to see how much a human can take. I've been denied a chance to not be in a wheelchair."

His views of his body image were varied and contradictory. He stated that

he was not a short person but a normal sized person with severely shortened arms and legs. Another time he stated, "I am short because my arms and legs dictate so." Still another time he stated, "I could live being a short person or a normal sized person but not both. I'm a new species. I'm more than a dwarf." He stated that he had never seen another person who looked like himself. He insisted that short or dwarfed women did not attract him as he did not have anything in common with them. If asked to draw himself, he would draw a man 5 feet tall. If hypothetically he were to become proportionate again, he stated that he would be less concerned about his hypertrochosis but he would attend to it eventually. He denied thinking that removing his hair would make him normal but he likened it to "fixing a fender on a wrecked car."

He was very guarded concerning his social history. He stated that he had a very unhappy childhood with much time spent in the hospital undergoing numerous surgical procedures on his club feet. He remembered his parents always arguing and described his mother as "cold and unloving." At some point when he was out of the hospital at 8 years old, he went home and 4 days later his mother left the family and moved far away. The next time he saw her was at age 23 when he lived with her for a while, but it did not work out because her new husband did not like him and at one point, came home drunk and beat him up. He had asked her why she left the family but she never answered him. He stated, "it's not necessary to know. She has her own life now." After age 8 he was raised by his father who he described as very supportive. He is the oldest of 1 sister, 1 half-brother, 2 half-sisters and 1 step-sister. He described his adolescence as "a living hell" where he was teased constantly in school. He claimed that he often physically fought back and won. He finished high school and worked intermittently as a bookkeeper while living with his father until about 4 years ago. He then lived in a Rehabilitation Center where he learned to become self sufficient and since that time has lived alone in an apartment. He at one time was active in The Little People of Quebec Society where he stated he saw mostly "midgets" with whom he felt he had nothing in common. He felt rejected by this group as he was primarily english-speaking whereas most of the members were french-speaking and insisted on conducting the meetings in french. He described his present relationship with his father and siblings as friendly but somewhat distant. "They're there if I need them," he stated. He related having very infrequent, distant interactions with his mother. He stated he never had a girl friend and felt extremely sexually isolated. He stated, "the girl that I love doesn't know I'm alive" but refused to elaborate any further. He said that he was not attracted to short women because he did not want a dwarfed child. "I would kill my child if he were a diastrophic dwarf," he stated.

#### PHYSICAL AND MENTAL STATUS EXAMINATION

The patient presented as a 3'10" male with grossly shortened limbs and a normal sized head and trunk. Examination of his nose was clear of hair growth as he had shaved there recently. These was a moderate amount of hair growth

evenly distributed on his back which was clearly within normal limits. He appeared to be moderately anxious and was easily moved to some sadness, but did not allow himself to cry. His speech was somewhat high pitched and irritating in quality. He was friendly, outgoing and displayed a sense of humor which was often self deprecating. He did not have any other symptoms of an affective disorder and there was no evidence of organicity or psychosis. He was very eager to talk with the interviewer but was also somewhat suspicious.

#### CLINICAL COURSE

The patient returned for several weekly dermatology visits as well as a psychiatric evaluation over the course of 3 months. He was instructed to not shave his nose and eventually a small number of hairs were observed which were treated by electro-epilation to his satisfaction. The psychiatric evaluation consisted of a single session and included a supportive psychotherapy emphasis and an attempt was made to continue this treatment during his dermatology visits. Gradually he gave up his concern for the disproportionate hair on his back after ventilating his frustration over the disappointment with his surgeon. He was repeatedly reassured that his back appeared normal and that many normal men have slightly uneven hair patterns on their back which are normal. He gradually accepted this and discontinued the dermatology visits with the understanding that he could return at any time in the future. Throughout the course of his treatment he remained resistant to speaking about his childhood and traumatic social experiences. When it was suggested that he may benefit from ongoing psychotherapy he adamantly refused stating that he did not need a therapist and that he controlled his own fate and was his own "psychoanalyst." He stated that he didn't want "to open up old wounds" but that he wanted to move on. One year after treatment, he was completely asymptomatic and deeply immersed in his college studies in political science with the goal of becoming the prime minister to "lead the handicapped people out of the woodwork."

#### DISCUSSION

The patient's story was unusual for patients with dysmorphophobia in that he presented with both a minimal defect—the hair on his nose—and a completely imagined deformity—the excessive and disproportionate hair on his back. The hair on his nose was diagnosed as trichostasis spinulosa—a benign condition treated by depilatory agents or electro-epilation. As far as his perception of his hairs on his back was concerned, he was felt to be within normal limits.

On the other hand, his distress over the ugliness of the "excessive and disproportionate" hair on his back was thought to be an obsession or overvalued idea. Although the belief occasionally took precedence over all other ideas, it did not have the false unshakeable quality of a belief of morbid origin characteristic

of a delusion. His complaint seemed to be less a conviction than a fear. While he expressed the belief that his excessive and disproportionate hair was responsible for his difficulty attracting women, he had also stated an awareness that it was not the sole cause. This was clear when he stated, "I know I am only fixing a fender on a wrecked car." He was also engaging and capable of forming a therapeutic relationship in the context of the dermatology setting.

In a tentative formulation, the precipitating event in this patient's illness was his perceived abandonment by his surgeon. This forced him to confront his intolerable dwarfed condition and to give up his deeply cherished hope to be normal sized. This event added to his already significant repressed hostility and covert self-reproach in a similar manner to his mother's abandonment of the family for which he may have blamed himself.

The early maternal rejection he suffered possibly during a period of oedipal striving may be at the root of his inability to accept his dwarfed condition. Perhaps the defective arms and legs were, in a sense, equated with defective genitals and thus were unconsciously taken as evidence of actual castration. At the same time that his physical condition deteriorated to requiring a wheelchair, he became more extreme in seeking a magical cure through surgery. His denial of his dwarfed condition convinced him that it was simply a matter of amputating his hands and feet and replacing them with longer prostheses to restore his body to normal. His wish for amputation may have been a regression to a masochistic, passive transference towards the surgeon. On the other hand, the final surgical result—longer, more normal looking prosthetic arms and legs—may have represented an undoing of his castration.

His condition may also be appreciated as a delayed developmental phenomena. As he presently existed, he felt alone claiming he had never seen another person like himself and indeed he had never found a peer group even among other little people. By becoming an amputee he would finally have a peer group thereby achieving an adolescent milestone and entering a larger community of disabled persons. He would no longer appear to others to have a congenital defect but instead one from an accident or trauma. He would be seen more as hapless victim than a mutation for which he might have found more tolerance as well as justification (consciously or unconsciously) for passivity and dependence.

The onset of his symptoms of hypertrichosis can be conceived as a regression in the ego function of reality testing. His somatic complaint of "disproportionate and excessive" hair demonstrates the use of displacement allowing him to keep the unpleasant reality of his dwarfism out of his conscious awareness in exchange for a more acceptable and treatable condition. The "disproportionate" quality of his hair is a symbolization for his disproportionate body and protects him from the anxiety attached to that intolerable condition. His negative feelings of bereavement and self-reproach are more acceptable as a physical symptom than as a direct discharge of emotional pain. Again removal of the disproportionate hairs may represent an unconscious undoing of his castration.



One year later when he was asymptomatic, immersed in school with dreams of political success, he had to some degree sublimated his wish to be a normal sized man with a more acceptable wish for academic and professional achievement. However, his grandiose visions of becoming a "big shot" may have been a reaction formation to his persistently low self esteem related to his small stature. Sometimes with physically disabled people the fear of passivity plays a part in spurring a person to greater endeavors as was shown in Lussier's analysis of a 13-year-old boy born with abnormally short arms who went on to become an accomplished trumpet player (30).

His resistance to expressive psychotherapy may be related to his fears of dependency and regression since he has experienced so much abandonment (real and perceived) in his life by parental figures. By presenting to dermatology he communicated what form of treatment he wanted. Because he did have a minimal defect—the hair on his nose—that could receive somatic treatment, he was able to feel comfortable with the combined dermatology—psychiatry approach. The supportive psychotherapy undertaken by both the psychiatrist and the dermatologist had the goals of bolstering his self esteem, allowing him to ventilate his frustration and strengthening his reality testing by reassuring him that his back looked normal.

The diagnosis of dysmorphophobia will need time for judgement to pass on it. It has only recently been elevated to a discrete illness by Thomas (22) and Hay (23) who have characterized it as an overvalued idea applying to patients whose only psychiatric problems are personality based such as schizoid, narcissistic or obsessional personality traits. It may be contrasted to monosymptomatic hypochondriacal psychosis (MHP), which Monroe (24) has defined as a single hypochondriacal delusion in an otherwise intact personality usually involving a distortion of body image. MHP is a subtype of paranoia classified in *DSM-III-R* as delusional (Paranoid) disorder, somatic type (297.10). The distinction between dysmorphophobia and MHP has been likened to the difference between a dysmorphic neurosis and a dysmorphic delusion respectively (25).

The treatment for dysmorphophobia includes insight or supportive psychotherapy and tricyclic antidepressants only if depressive features are pronounced (3). Patients with this disorder tend to be more mature, capable of forming a therapeutic relationship and capable of varying degrees of insight.

MHP had been reputed to respond specifically to pimozide (26,27) in some early uncontrolled, noncomparative studies. Haloperidol (3), fluphenazine decanoate (31), tricyclic antidepressants (28), and MAOI's (29) have since also been found to be effective in case reports. Patients with MHP are usually not good candidates for psychotherapy as they are often immature, incapable of insight and incapable of forming a therapeutic relationship (3). When the diagnosis remains unclear between the two dysmorphic disorders a diagnostic-therapeutic drug trial may be indicated (22). Because these two disorders have only been recently clearly distinguished from each other there is no clear data to determine long-term psychological outcome.

In summary, there is an unclear relationship between patients who have real defects versus psychological defects in their body image. As this case illustrated, sometimes a psychological defect can displace and symbolize an intolerable real defect when denial becomes unbearable. Also, it is suspected but not clearly shown that dwarfs have an increased amount of psychological difficulties. Diastrophic dwarfs may be particularly predisposed to psychiatric problems because of their inevitable morbidity involving limitations of mobility but to date no studies have been done to confirm this. Finally, although the long-term psychological outcomes of the dysmorphic disorders remain unclear, this case illustrates an effective treatment of dysmorphophobia by an innovative psychiatry-dermatology liaison approach of supportive psychotherapy and somatic treatment.

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