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Uncertainty and Posttraumatic Stress: Differences between Mothers and Fathers of Infants with Disorders of Sex Development

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Abstract

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Ethical Approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

Conflicts of Interest: Dr. Paul Austin - Pediatric advisory group & clinical investigator for Allergan and Dr. Dix Poppas - Co-founder of Promethean Surgical Devices, Inc. No other conflicts of interest.

Parents of children with Disorders of Sex Development (DSD) report significant psychological distress, including posttraumatic stress symptoms (PTSS), with mothers consistently reporting higher rates of psychological distress than fathers. However, psychological factors contributing to PTSS in both parents are not well understood. The present study sought to fill this gap in knowledge by examining PTSS and illness uncertainty, a known predictor of psychological distress, in parents of children recently diagnosed with DSD. Participants were 52 mothers (M_{age} = 32.55 years, SD = 5.08) and 41 fathers (M_{age} = 35.53 years, SD = 6.78) of 53 infants (M_{age} = 9.09 months, SD = 6.19) with DSD and associated atypical genital development. Participants were recruited as part of a larger, multisite study assessing parents' psychosocial response to their child's diagnosis of DSD. Parents completed measures of illness uncertainty and PTSS. Mothers reported significantly greater levels of PTSS, but not illness uncertainty, than fathers, and were more likely than fathers to report clinical levels of PTSS (21.2% compared to 7.3%). Hierarchical regression revealed that parent sex, undiagnosed or unclassified DSD status, and illness uncertainty were each associated with PTSS. The overall model accounted for 23.5% of the variance associated with PTSS. Interventions targeting illness uncertainty may be beneficial for parents of children with newly diagnosed DSD.

Keywords

Disorders of Sex Development; DSD; Intersex; Posttraumatic Stress; Illness Uncertainty

INTRODUCTION

Disorders of sex development (DSD) are congenital conditions that are characterized by discordance between aspects of biological sex, such as sex chromosomes and internal/external sex structures (Hughes, Houk, Ahmed, & Lee, 2006). Infants born with DSD may undergo complex, long-term medical and surgical management, and some forms of DSD are life-threatening and/or may be associated with increased cancer risk (Clayton, Miller, Oberfield, & Sippell, 2002; Kim & Kim, 2012). For infants born with DSD and associated atypical genital development, gender designation at birth can be challenging and contribute to uncertainty for parents (Hughes et al., 2006; Kolesinska et al., 2014). Approximately 50% of affected individuals receive a diagnosis of an underlying etiology when there is the presence of a Y chromosome (Barseghyan, Délot, & Vilain, 2015), and guidelines for gender of rearing are less certain when a diagnosis is lacking (Bakula et al., 2017; Kolesinska et al., 2014). In addition to facing the psychological stress of adjusting to having a child diagnosed with a chronic condition, parents may be presented with a host of medical decisions about the management of their child's DSD in the first months and years after birth. This decision-making includes whether or not to proceed with surgery (e.g., genitoplasty, gonadectomy), depending on the etiology and presentation of their child's DSD (Mouriquand et al., 2016). Parents often report considerable psychological distress surrounding these decisions (Brain et al., 2010; Kirk et al., 2011; Wiesemann, Ude-Koeller, Sinnecker, & Thyen, 2010).

The American Psychiatric Association (APA) previously asserted that knowledge of a child's life-threatening illness may be considered a traumatic event (APA, 2000). Notably, parents of children with DSD report posttraumatic stress symptoms (PTSS) at rates similar

to parents of children with life-threatening cancers, regardless of whether or not their child's DSD has the potential to be fatal (Duguid et al., 2007; Pasterski, Mastroyannopoulou, Wright, Zucker, & Hughes, 2014; Suorsa et al., 2015). Pasterski et al. conceptualized the experience of having a child diagnosed with DSD within the context of the Pediatric Psychosocial Preventative Health Model (Kazak et al., 2006). This is a model widely used in pediatric oncology to understand the idiopathic nature of PTSS, as it posits that it is not only the event (e.g., diagnosis), but also the interpretation of the event that contributes to PTSS. For parents, many events following their child's diagnosis of DSD have the potential to be viewed as traumatic, including the process of gender designation, surgery, and how others respond to knowledge of the child's condition (Pasterski et al., 2014).

Parental distress levels vary by parent sex and child gender designation (Pasterski et al., 2014; Slijper, Frets, Boehmer, Drop, & Niermeijer, 2000; Suorsa et al., 2015; Wolfe-Christensen et al., 2012; Wolfe-Christensen, Fedele, Mullins, Lakshmanan, & Wisniewski, 2014). Previous findings from our multisite group, including a subsample of the cohort presented in this article, show that parents of boys with DSD report higher rates of illness uncertainty than parents of girls shortly after their child's diagnosis (Suorsa et al., 2015). Further, other research suggests that parent sex differences and child gender of rearing are important to consider in this population. For example, mothers of children with DSD are more likely to endorse clinical symptoms of PTSD than fathers (Pasterski et al., 2014), as well as other symptoms of distress, such as anxiety (Wolfe-Christensen et al., 2014). For caregivers of boys, parental psychological distress has been related to degree of undervirilization of their child's genitalia at birth (Wolfe-Christensen et al., 2012). Mothers also report negative adjustment outcomes over a more extended period of time than fathers (Slijper et al., 2000). Understanding of the factors contributing to these sex differences among parents is limited, in part due to global underrepresentation of fathers in research studies (Phares, Fields, Kamboukos, & Lopez, 2005; Phares, Lopez, Fields, Kamboukos, & Duhig, 2005). Given that parenting a child with DSD has the potential to result in negative adjustment outcomes, further examination into variables that may contribute to distress is warranted for both mothers and fathers.

Parental illness uncertainty is a cognitive appraisal variable characterized by the perceived lack of information, or clarity, about a child's condition and its accompanying treatment (Mishel, 1983; Stewart & Mishel, 2000). It has been associated with PTSS for mothers of children with a number of chronic illnesses, including diabetes, rheumatic diseases, and cancer (Fedele et al., 2011; Fuemmeler, Mullins, & Marx, 2001; Hoff, Mullins, Chaney, Hartman, & Domek, 2002; Tackett et al., 2015). Though the extant findings are limited, illness uncertainty may be an important contributor to distress for parents of children with DSD as well. Earlier studies from our group reported that greater levels of illness uncertainty at presentation were related to later decisional regret regarding medical decisions made for their child with DSD (Ellens et al., 2017). Pasterski et al. (2014) reported that for parents of children with DSD participating in a cross-sectional analysis, increased "cognitive confusion" about their child's condition was associated with PTSS. Similarly, Crissman et al. (2011) found that uncertainty about their child's DSD diagnosis and medical management was related to parents' stress levels. Recent studies, including our own, have also found that parents of children with DSD report levels of illness uncertainty and PTSS

that are comparable to those reported by parents of children with cancer (Pasterski et al., 2014; Suorsa et al., 2015), for whom uncertainty is a known predictor of PTSS. Despite high rates of both illness uncertainty and PTSS in parents of children with DSD and an established relationship between these variables in other chronic conditions, such associations have not been fully examined within the context of having a child with DSD.

As such, the present study sought to expand upon previous findings regarding illness uncertainty, PTSS, and parental sex differences in a cross-sectional sample of parents of children with DSD (i.e., Pasterski et al., 2014). To do so, this study utilized a prospective sample of parents to better understand initial psychosocial responses to having a child born with DSD and associated atypical genital development. The present study had three primary aims: (1) to examine differences on ratings of illness uncertainty and PTSS for mothers and fathers, (2) to examine differences on ratings of illness uncertainty and PTSS for parents of boys compared to parents of girls and parents of infants with an identified specific diagnosis of DSD compared to parents of infants with undiagnosed/unclassified DSD, and (3) to explore the relation between uncertainty and PTSS for mothers and fathers. We hypothesized that mothers, parents of boys, and parents of children with an undiagnosed/unclassified DSD would report the greatest levels of illness uncertainty and PTSS. Additionally, we expected that higher rates of uncertainty would be related to higher rates of PTSS.

METHOD

Participants

Participants were 52 mothers and 41 fathers of 53 infants with DSD and associated atypical genital development. Degree of atypical genital development was classified as a Prader rating of 3 to 5 in cases of 46,XX DSD and a Quigley rating of 3 to 6 in cases of DSD where a Y chromosome was present (Prader, 1954; Quigley et al., 1995). Inclusion criteria comprised having a child less than 24 months of age. There were 40 parental dyads, meaning that for 40 of the infants in the study, both parents participated. An additional 13 parents completed the study on their own. Participants were predominantly White (67.6%). Of the remaining participants, 6.4% were Black/African American, 7.5% were Asian/Pacific Islander, 4.3% were multiracial, 6.4% identified as “other race,” and 7.5% declined to identify their race. Of the overall sample, 22.6% also identified as Hispanic. Most parents reported that they were married to the other parent of their child with DSD (83.3%). Thirty children were affected by 46,XX DSD, 18 children by 46,XY DSD, and 5 children by sex chromosome DSD. All infants with 46,XX DSD were raised female. Sixteen infants with 46,XY DSD were raised male and 2 were raised female. Three infants with sex chromosome DSD were raised male and the parents of the other 2 infants reported being unsure/undecided of their child’s gender designation at the time of participation. Parents of 11 infants with 46,XY DSD and 2 infants with sex chromosome DSD had not received a diagnosis of an underlying etiology of their child’s DSD; they all reported raising their child male. None of our families had more than one child with DSD at the time of participation. Informed consent was obtained from all individual participants included in the study. Descriptive statistics are shown in Table 1.

Measures

Demographics—The Demographics Form consisted of items assessing parent-reported child gender of rearing, child age and date of birth, DSD diagnosis, parent relationship to child (e.g., biological mother or father), parent marital status, parent race/ethnicity, parent date of birth, and others.

Parent Perception of Uncertainty Scale (PPUS)—The PPUS (Mishel, 1983) is a 31-item self-report questionnaire that measures a parent's perception of illness-related uncertainty surrounding their child's health problem. Item responses are presented on a 5-point Likert scale ranging from *Strongly Agree* to *Strongly Disagree*. Scores range from 31 to 155. Higher overall scores on the PPUS indicate higher levels of uncertainty. An example of an item on the PPUS is, "My child's treatment is too complex to figure out." In addition to an overall score, which was utilized for the present study, the PPUS consists of four factor scores. These factors are: Ambiguity, Lack of Clarity, Lack of Information, and Unpredictability. The PPUS is a well-established measure of parental illness uncertainty in the area of childhood chronic illness, evidencing moderate to high coefficient alphas (α = .86 to .93; Mishel & Epstein, 1997). The total PPUS yielded high internal consistency for both mothers (α = .89) and fathers (α = .93) for this sample.

Impact of Events Scale-Revised (IES-R)—The IES-R (Weiss & Marmar, 1997) is a 22-item self-report measure widely used to assess PTSS. Respondents are asked to rate their feelings related to a potentially traumatic event, in this case their child's diagnosis of DSD (e.g., "Any reminder brought back feelings about it.") on a 5-point Likert scale, ranging from *Not at All* to *Extremely*. Higher scores indicate an increased amount of PTSS. A total score of 33 or greater on the IES-R indicates clinically significant PTSS, and would likely meet criteria for a diagnosis of posttraumatic stress disorder (PTSD; Weiss & Marmar, 1997). In addition to an overall score, which was utilized for the present study, the IES-R provides three subscale scores: Avoidance, Hyperarousal, and Intrusion. Using two separate standardization samples, Weiss and Marmar reported coefficient alpha's ranging from .79 to .92 for the three subscales. The total IES-R yielded high internal consistency for both mothers (α = .95) and fathers (α = .93) for this sample.

Procedure

Institutional review board approval was obtained from participating institutions. Parents were recruited from 11 clinic sites specializing in DSD care from around the United States. They were approached during regularly scheduled clinic appointments. Parents were enrolled as part of a larger prospective study assessing psychosocial factors related to a new DSD diagnosis. All parents were eligible to participate regardless of their decision to proceed, or not, with various types of medical or surgical treatment for their child. Data presented here were collected prior to genitoplasty if parents chose to have that surgery for their child. Families were compensated for travel costs to the center.

Statistical Analyses

Statistical analyses were conducted using SPSS® version 23. Listwise deletion was used for missing data. To characterize our sample, the percentage of parents who reported clinically

significant PTSS, based on the established cutoff score of 33 on the IES-R, was examined. To account for the relationship between mother-father dyads, a mixed between-within subjects analysis of variance (ANOVA) was used to examine the effects of parent sex, child gender of rearing, and presence of diagnosis on illness uncertainty and PTSS. Bivariate correlations were then conducted to explore the relations between variables of interest, including overall illness uncertainty, level of PTSS, DSD karyotype, and degree of genital atypicality (i.e., Prader and Quigley ratings), and child age. Hierarchical regression was then used to assess the effect of parent sex, diagnostic status (i.e., whether or not an affected child had/had not received an etiological diagnosis of their DSD), and perceived illness uncertainty on PTSS. Prior to conducting the regression analyses, preliminary tests were employed to ensure no violation of the assumptions of normality, linearity, multicollinearity, or homoscedasticity occurred. For this analysis, bias-corrected bootstrapped resampling with replacement was used at the 95% confidence interval level, specified for 5,000 redraws (Hayes 2013). A significance threshold of .05 was utilized.

RESULTS

Of parents who completed the IES-R, 13.5% met or exceeded the cutoff score for a clinically significant level of PTSS. Specifically, 21.2% of mothers and 7.3% of fathers met or exceeded this threshold. ANOVA revealed no significant interaction between, or main effect for, parent sex, child gender of rearing, or diagnostic status on illness uncertainty ($p > .05$). However, there was a significant within-subjects main effect of parent sex on PTSS, Wilks' lambda = .71, $F(1, 33) = 13.62$, $p = .001$, and a significant between subjects effect of diagnostic status on PTSS, $F(1, 33) = 6.79$, $p = .014$. Mothers reported greater PTSS than fathers and parents of infants who were in the undiagnosed/unclassified DSD group reported higher PTSS than parents of infants with a diagnosis. The interactions between parent gender and diagnostic status and child gender were not significant ($p > .05$). Mean scores from the ANOVA analyses are shown in Table 2.

Bivariate correlations revealed that degree of child genital atypicality, DSD karyotype, and child age were not correlated to outcomes variables for this sample; thus, these variables were not included in the regression model. Due to significant differences revealed by the ANOVA, parent sex and diagnostic status were included as predictors in the final regression model examining the association between illness uncertainty and PTSS. Parent sex and diagnostic status were entered on Step 1 of the hierarchical regression, and accounted for 14.8% of the variance in PTSS. Illness uncertainty was entered on Step 2, accounting for an additional 8.7% of the variance in PTSS, after controlling for parent sex and diagnostic status, R^2 change = .087, F change (1, 82) = 9.36, $p = .003$. The total variance accounted for by the model as a whole was 23.5%, $F(3, 85) = 8.39$, $p < .001$, $R = .048$, $R^2 = .24$. Parent sex, diagnostic status, and degree of illness uncertainty all significantly contributed to the final model ($p < .05$). Results from the regression analysis are shown in Table 3.

DISCUSSION

The current study examined illness uncertainty and PTSS in parents of infants newly diagnosed with DSD and secondary atypical genital development. Our findings indicated

that a subset of parents viewed their infant's diagnosis as a traumatic event. Mothers were particularly vulnerable to the development of PTSS, as they were almost three times more likely to report clinical levels of PTSS than fathers. Such results are consistent with previous research conducted with a subsample of this cohort (Suorsa et al., 2015) in addition to other independent research (Pasterski et al., 2014). Although it is unclear why mothers demonstrate higher levels of PTSS than fathers, Wolfe-Christensen et al. (2014) hypothesized that fathers' relatively lower levels of distress may be due to the use of disengagement to cope with their child's illness. Research supports this hypothesis, suggesting that men and women utilize different coping styles when faced with a stressful life experience, such as having a child diagnosed with a chronic illness (Chaney et al., 1997; Mastroyannopoulou, Stallard, Lewis, & Lenton, 1997; Tamres, Janicki, &Helgeson, 2002).

Both mothers and fathers in our sample were nearly twice as likely to report clinical PTSS as would be expected from lifetime prevalence data in the general population (Kessler, Chiu, Demler, Merikangas, & Walters, 2005). Previous findings suggest rates of clinical PTSS as high as 31% for mothers and 18% for fathers of children with DSD (Pasterski et al., 2014). Our results are also consistent with research conducted by our group and others suggesting that parents of children with DSD experience rates of PTSS at levels similar to those reported by parents of children with cancer (Pasterski et al., 2014; Suorsa et al., 2015). Additionally, analyses revealed that parents of children whose DSD was undiagnosed or unclassified, all of whom were reared boys, reported greater levels of PTSS than parents of infants with DSD for whom an underlying etiology was identified. This is consistent with previous findings that parents of boys with DSD demonstrate greater psychological distress than parents of girls (e.g., Wolfe-Christensen et al., 2012). Furthermore, our results build on this finding by suggesting that parents of boys may report greater distress due to the lack of a clear diagnosis for their children. Our findings also suggest that illness uncertainty is an important contributor to PTSS for parents of infants with DSD, despite diagnostic status. When controlling for parent sex and child diagnostic status, illness uncertainty accounted for a significant amount of the variance associated with PTSS. The overall model indicated a medium to large effect (Meyer et al., 2001), and revealed parent sex, undiagnosed/unclassified DSD status, and illness uncertainty all to be salient predictors of PTSS. These results are similar to other research conducted with parents of children with chronic illnesses, such as cancer, finding illness uncertainty to be related to PTSS (e.g., Tackett et al., 2015).

Additionally, in a cross-sectional sample of parents representing a broad range of time since diagnosis, Pasterski et al. (2014) found cognitive confusion (ostensibly a proxy for uncertainty) to be a driving factor behind PTSS in parents of children with DSD, regardless of parent gender. Our findings build upon the Pasterski et al. study by examining the relationship between illness uncertainty and PTSS. Cumulatively, such evidence suggests that uncertainty, although not the only factor associated with PTSS, is indeed significantly related to PTSS in parents of children with DSD. As previously reported, parents of children with DSD often lack information related to their child's condition, including information about future outcomes for their child (Duguid et al., 2007), even when they have received an etiological diagnosis of their child's DSD.

Implications

Findings from the present study revealed that some mothers and fathers experienced illness uncertainty and PTSS shortly after their child's diagnosis, the same time frame within which they are engaging in medical decision-making for their child. As indicated by previous research on medical decision-making in parents of children with chronic illnesses (Pyke-Grimm, Stewart, Kelly, & Degner, 2006), this level of uncertainty and psychological distress may impact parents' perceptions of their ability to make medical decisions for their child. Initial analyses conducted with a subset of the current sample indicated a downward trend in PTSS for both mothers and fathers by 12-month follow-up (Ellens et al., 2017), and previous research in mothers of children with cancer also supports this downward trajectory (Dolgin et al., 2007), suggesting that overall parents adjust to their child's diagnosis over time. However, our group also showed that a greater degree of illness uncertainty reported at baseline was related to increased regret surrounding decisions about their child with DSD at 12-month follow-up (Ellens et al., 2017). Indeed, it appears that a subset of parents report distress regarding medical decision-making (e.g., gender designation and/or genitoplasty decisions) for their child over time (Brain et al., 2010; Kirk et al., 2011; Wiesemann et al., 2010). Future research should examine variables, including cognitive appraisals, impacting decision-making in parents to better understand parents' initial and ongoing need for psychological intervention and support.

Greater information regarding the experience of uncertainty is needed in this population, as well. Interestingly, lack of a diagnosis in parents of children with DSD in which a Y chromosome was present was not related to parents' rating of illness uncertainty, although it was significantly associated with levels of PTSS. Diagnostic information is certainly invaluable for parents of children with DSD when engaging in decision-making; and the process by which parents obtain genetic testing results will be an important ongoing area of improvement as technology progresses and becomes more available (Kutney, Konczal, Kaminski, & Uli, 2016). However, lacking a diagnosis was not the only factor related to PTSS for our families. Parent sex and overall illness uncertainty were also significantly associated with PTSS, and these findings could be used to inform psychological supports. Specifically, interventions targeting illness uncertainty by teaching problem solving and cognitive reframing skills may be beneficial, as they have been shown successful in parents of children with other chronic conditions (Hoff et al., 2005; Mullins et al., 2012). In addition, further research examining specific contributors to uncertainty, as well as the trajectory of distress, in parents of children with newly diagnosed DSD will provide much needed information to inform improved psychological support systems for this population.

Results from the present study provide insight into the psychological functioning of both mothers and fathers of infants newly diagnosed with DSD including atypical genital development. However, these findings are not without limitations. First, this study reports data from a single time point. As such, the directionality between the association of parental uncertainty and PTSS cannot be concluded. Second, our study was limited in its ability to provide information regarding factors influencing illness uncertainty and distress in this population. Third, we do not have consistent data regarding who chose to participate in our study, as rate of participation and reason for not participating was not consistently tracked

across each site involved in the study. As such we cannot accurately infer the generalizability of our findings. It may be that parents with the most extreme emotional responses to their child's diagnosis elected to participate.

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Table 1.

Descriptive Statistics

Age	***N***	***M* (*SD*)**
Child Age	53	9.09 months (6.19)
Mother Age	52	32.55 years (5.08)
Father Age	41	35.53 years (6.78)
Diagnosis by Karyotope	***N***	**% Karyotype, % Overall**
46,XX DSD	**30**	
Congenital adrenal hyperplasia	28	93.3, 52.8
Other	2	6.7, 3.7
46,XY DSD	**18**	
46,XY with hypovirilization unknown cause	11	61.1, 20.7
Androgen insensitivity syndrome	2	11.1, 3.7
Gonadal dysgenesis	3	16.7, 5.6
Other	2	11.1, 3.7
Sex Chromosome DSD	**5**	
45,XO/XY	3	60, 5.6
Other/Unclassified	2	40, 3.7
Gender of Rearing by Karyotype	***N***	**% Karyotype, % Overall**
46,XX DSD		
Girl	30	100, 56.7
Boy	0	0, 0
Undecided	0	0, 0
46,XY DSD		
Girl	2	11.1, 3.7
Boy	16	88.9, 30.2
Undecided	0	0, 0
Sex Chromosome DSD		
Girl	0	0, 0
Boy	3	0, 0
Undecided	2	40, 3.7

Note. *M* = mean score. *SD* = Standard deviation

Table 2.

Mean Scores for ANOVA Variables

	PPUS[a] *M* (*SD*)[c]	IES-R[b] *M* (*SD*)
Mothers of girls	**62.09 (16.65)**	**19.55 (17.33)**
Diagnosed	62.09 (16.65)	19.55 (17.33)
Undiagnosed[d]	--	--
Mothers of boys	**72.31 (12.93)**	**30.00 (19.89)**
Diagnosed	73.00 (13.02)	14.75 (10.99)
Undiagnosed	72.00(13.67)	36.1 (19.66)
Fathers of girls	**61.83 (15.49)**	**8.77 (9.63)**
Diagnosed	61.83 (15.49)	8.77 (9.63)
Undiagnosed[d]	--	--
Fathers of boys	**70.93 (20.88)**	**17.36 (15.87)**
Diagnosed	71.50 (8.10)	6.00 (3.56)
Undiagnosed	70.67 (25.08)	21.90 (16.71)

[a] PPUS = *Parental Perception of Uncertainty Scale*. Total scores for the PPUS range from 31 to 155.

[b] IES-R = *Impact of Events Scale-Revised*. Total scores for the IES-R range from 0 to 80.

[C] *M* = mean score. *SD* = Standard deviation.

[d] All infants who were reported to be raised as girls received a diagnosis.

Table 3.

Regression Analysis[a]

	β	t	p
Step One			
Parent Sex	−.28	−2.75	.007
Diagnostic Status	.27	2.74	.007
Step Two			
Parent Sex	−.27	−2.83	.006
Diagnostic Status	.24	2.47	.016
Illness Uncertainty	.29	3.05	.003

[a]Outcome variable = Impact of Events Scale-Revised