An Altered Life Process: Tourette Syndrome

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Abstract

Tourette Syndrome (TS) is one of the rarest forms of neurologic disorders that affect children. The precise etiology of TS is unknown, but the primary cause of this disorder is a "developmental disorder of synaptic neurotransmitter reuptake in the cortico-straital-thamlamic circuitry of the brain" (Rindner, 2007, p. 20). The secondary cause of TS is associated with the following risk factors: hereditary factors such as the autosomal linked disorder; repeated streptococcal infections in early life in children with preexisting health conditions; and comorbidities such as depression, anxiety, obsessive compulsive disorder and attention deficit hypersensitivity disorder. These comorbidities make the diagnosis of TS complex. TS affects the mental, physical, emotional, and psychosocial well-being of a client. TS cannot be fully cured, but it can be controlled by using pharmacological and non-pharmacological interventions. Nurses play the important role of advocate for the client. Family centered care, social support, active listening, and empathetic communication are qualities of an effective therapeutic intervention for clients suffering from TS and their families.

Understanding Tourette Syndrome: Pathophysiology

Tourette Syndrome (TS) is one of the rarest forms of neurologic disorder, affecting no more than 10 of every 1,000 (Scahill, et al., 1993). TS is observed in the beginning childhood years and is usually characterized by numerous autonomic physiological movements called "motor tics" and uncontrollable vocal movements called the "phonic" or "vocal tics" (Centers for Disease Control, 2010).

The exact etiology of TS is unknown. However, research indicates that about 60% of cases seen in children under the age of 18 exhibit a hereditary link that manifests some autosomal dominant pattern. However, the research cannot rule out possibilities of multifactorial or intermediate mode of inheritance for TS (Schapiro, 2002). Golder's study suggests that in younger children, decreased dopamine in the brain activates the development of tics (Golder, 2010). Multiple epidemiological, genetic, neuroimaging, and postmortem studies conducted by Jankobic (2001) indicate that TS is an "inherited developmental disorder of synaptic neurotransmitter reuptake in the cortico-straital-thamlamic circuitry of the brain" (as cited in Rindner, 2007, p. 20). The basal ganglion, the primary center of the brain involved in motor control, cognition, and emotions, is associated with the pathogenesis of this neurologic disorder (Rindner, 2007). Several studies of children with TS have reported increased risk for TS of 11.5% in brothers and 4.8% in sisters (Tourette Syndrome Association, 1999). Research has also suggested that an "autosomal dominant model with sex-specific markers" results in greater TS prevalence among the male population, with males having four times greater incidence (Golder, 2010).

Another pathophysiological factor associated with TS is streptococcal infections in children in early life; these have been identified as "pediatric autoimmune neuropsychiatric disorders associated with streptococcal infections (PANDAS)" (Cavanna et al., 2009, pp. 13-23). This phenomenon is

especially observed in the pediatric population under the age of 11 (Golder, 2010).The streptococcal infection reduces the brain activity in the basal ganglia by decreasing the production and use of dopamine (Cavanna et al., 2009). As a result, children exposed to streptococcus infection may experience an abrupt onset of motor tics (Golder, 2010).

The pathophysiology of TS also includes comorbidity with other psychological disorders. Ninety percent of all children diagnosed with TS exhibit more than one comorbid disorder. A study by Golder (2010) indicates that more than half of the children diagnosed with TS are also affected by anxiety, obsessive-compulsive disorder (OCD) and behavioral problems (as cited in Jimenez, 2008). Similarly, other comorbidities related to TS include mood disorders such as depression, which occurs in 10% of all known TS patients (Cavanna et al., 2009).

It is difficult to make a diagnostic test for TS due to its unknown etiology. However, functional symptoms can be examined and assessed to distinguish the syndrome from other illnesses or conditions that cause involuntary tics (Gelbart, 1998).

Bagheri & Leckman (1999) provide a thorough explanation about the types of tics and their symptoms that demonstrate acute physical exacerbation, as in TS (as cited in Schapiro, 2002). Tics in TS generally start between the ages of three to eight years (Jimenez, 1999). Studies by Bagheri & Leckman (1999) indicate that simple motor tics include "shoulder shrugging, neck twitching, and facial movements, as well as feeling objects, hopping, and bending in sporadic fashion; likewise, behaviors such as sniffing, barking, coughing, yelling, and hiccupping" are considered as simple vocal tics; complex vocal tics include behaviors such as reiteration of words, phenomenon of self talking, and using profane and inappropriate words (as cited in Schapiro, 2002, p. 243).

Pharmacological and Non-pharmacological Treatments

The complex neurologic disorder Tourette Syndrome (TS) involves imbalances "of synaptic neurotransmitter reuptake in the cortico-stratial-thalmic circuitry of the brain"; the pathogenesis of the disorder becomes difficult to understand due to its complexity and association with other comorbidities such as "obsessive compulsive disorder (OCD), attention deficit hyperactivity disorder (ADHD), anxiety" and depression (Rindner, 2007, p. 20). Similarly, the intrinsic interaction among genes and the numerous physiological manifestations such as neck jerks, shoulder shrugs, and abdominal tensing also contribute to health risks, making the clinical study of TS even more challenging (Rindner, 2007). Numerous people with TS who endure mild tics do not require pharmacological assistance. However, when "tics" start to interfere with everyday life, medication is used to control or to minimize the symptoms (Ohm, 2006).

The most useful drugs that have demonstrated effectiveness in controlling tics are the typical neuroleptic agents such as haloperidol (Haldol) and primozide (Orap). Haldol was the only drug of choice for TS during the 1960s and '70s. Research indicates that Haldol and Orap, the antidopamindergic drugs, have produced reductions in tics between 34-67% among clients suffering from TS (Peterson & Azrin, 1993). However, due to its serious side effects and drug-drug interaction such as "tardive dyskinesia, dysphoria, akathisia and depression of the central nervous system" (CNS), Haldol is no longer used (Schapiro, 2002, p. 251).

Similarly, other atypical antipsychotic agents such as risperidone (Risperdal), olanzapine (Zyprexa), and ziprasidone (Geodon) have been used to control tics (Rindner, 2006). These drugs are commonly used in the treatment of TS due to lesser "side effects such as sedation and extrapyramidal symptoms" (Jimenez, 2009, pp. 738-739). Other categories of drugs such as alpha-2 beta agonist, which includes the drug clonidine, have been used with tics and ADHD symptoms (Rindner, 2007). However, clinical research indicates that the alpha 2 adrenergic agonist drugs inhibit the release of the noradrenaline hormones responsible for the flight and fight responses in the human body (Jimenez, 2009). Thus, it is important to realize that some of the aforementioned drugs used for the treatment of tics have adverse effects, altering psychosocial and physiological phenomena, causing weight gain, movement disorder, cardiovascular disorder, and metabolic disorder (Schapiro, 2002). The main purpose of providing these drugs to a client with TS is to control the long-term symptoms of tics as opposed to the short-term symptoms (Jimenez, 2009). Whenever possible, care providers recommend seeking non-pharmacological treatment to control the tics, due to the severe side effects of pharmacological drugs (Ohms, 2006).

Non-pharmacological therapy includes biofeedback or relaxation techniques to reduce stress, breathing techniques and cognitive-behavioral therapy to reduce anxiety and increase concentration,

and social skills training by involvement in support groups and social activities to develop friendships, build trust among family and friends and enjoy life (Ohms, 2006). Initially, clients with TS find it difficult to form habits and to perform tasks by applying the non-pharmacological interventions to reduce tics; however, research indicates that non-pharmacological intervention, particularly psychosocial treatment, is 55-95% more effective in terms of reducing the frequency of tics (Blacher, 2006).

Further research (Jimenez, 2009) indicates that treatments and management of severe TS include use of deep brain stimulation (DBS). The research indicates that effectiveness of DBS is seen among clients over 25 years of age who have severe or chronic tic disorder. A prospective randomized trial of DBS conducted on TS has shown positive correlation in treating severe tics by stimulating the frontal thalamus region of the brain; however, there is no clear evidence to indicate that DBS would be more effective than other treatments. Thus, further study is necessary to determine the true effectiveness of DBS, including its adverse effects (Jimenez, 2009).

Family Implications

Family centered care (FCC) is an important aspect of nursing, as it gives nurses opportunity to understand the client and his or her family as a complete unit. Family System Nursing allows nurses to explore family dynamics and provide appropriate care towards health promotion and disease prevention. For example, addressing the health needs of a child suffering from TS can affect the health of other family members, because they are directly or indirectly providing care to the child. Similarly, FCC nursing collaborates with members of the family to recognize the importance of interdependence among members of the family in attaining and in promoting health. FCC empowers family members to become active participants in the decision making process of their own health, as well as support and advocate for the overall well being of their family members (Bell, 2009).

Because TS is seen as a form of disability in a child (Lee et. al, 2007), the family may feel overwhelmed by the diagnosis and may have difficulty coping with the illness (Ohm, 2006). Nursing research indicates that the stress of taking care of a child with TS affects the caregiver's relationship with other family members (Kersh, 2006). When the family members are engaged and involved with the child with the disability, due to his/her physical mental and emotional needs, there is less time to take care of other children. Similarly, parents seem to experience strain in their relationship due to lack of communication with each other and less time for them to relax and enjoy their life. Due to this overwhelming feeling, parents sometimes refrain from getting involved in the care of their child (Kersh, 2006). Thus, the stress of caring for the child with illness demands that family members approach their own life in a meaningful and skillful way (Lee et. al, 2007).

In order to deal with the obstacles and challenges posed by the illness in the life of the client and his/her family members, parents should identify ways to maintain a healthy lifestyle. Identifying the stressors of life, such as the child's medical diagnoses and economic and emotional stressors caused by the illness, and availability of a support system outside the family, are important in order to maintain a safe and healthy life. Also, seeking activities such as psychosocial therapy is a constructive coping mechanism in regulating the healthy atmosphere of the family (Ohm, 2006). Similarly, getting involved in charitable or volunteer work, attending social meetings/support groups to share feelings with other families who are faced with similar challenges, and expressing emotions can help family members "empathize and identify with others" (Ohm, 2006, p. 195). These aforementioned coping skills can help decrease anxiety and reduce the stress.

Nursing Implications

Children and adolescents with Tourette Syndrome live a complex life because they are faced with uncertainty and challenges as a result of the uncontrollable motor and vocal tics which can suddenly occur in their behavior any time during the day (Rindner, 2007). They may experience difficulty trying to suppress the tics and may become nervous in the classroom setting (Golder, 2010). Due to the sudden onset, the children might face challenges concentrating in the classroom. They may also experience negative responses from their friends, such as bullying and/or isolation. Thus, the phenomenon of TS can cause mental, psychological and physical stress on children and adolescents (Golder, 2010). Therefore, as nurses, it is important to recognize and understand the typical onset of the tics, which includes sudden and unusual symptoms such as neck jerks, shoulder shrugs, abdominal tensing and so on. Understanding the manifestations of TS, recommending appropriate treatment methods, and serving

as referral sources for clients and their families are primary ways nurses can advocate for their client and family. Similarly, in educating the client, nurses should be aware of the social, educational, and physical challenges faced by youth. For example, being bullied by peers and strangers can impact the client's self esteem. Thus, special emotional support should be provided to these populations (Rindner, 2007).

Understanding the needs of the client and their family members begins by educating others about TS (Rindner, 2007). Nurses can advocate for their clients by educating individuals involved in the client's daily life about the treatment, coping skills, and emotional and physical trauma brought by TS. It is important to tell students, teachers, and supervisors that behavior exhibited by clients suffering from TS is not the client's fault and not something the client voluntarily chooses to do so. Likewise, nursing care should also depend upon individual needs. It will allow others to be nonjudgmental about the client's disease process, separate the client from the disease processes, and respect the client as a unique individual.

Research recommends that teachers provide individual attention to students with TS, to promote their effective education as well as communication in classroom settings (Rindner, 2007). One of the ways that teachers could contribute to the needs of students with TS could be by creating special arrangements for students suffering from tics. For example, allowing the child to move around in the classroom, offering an empty office or spaces for the children with tics to move around until the symptoms subside, and permitting them to use tape recorders in the classroom will alleviate academic anxiety and reduce the pressure on them (Castiglia, 2001). In this way, special sensitivity, empathetic consideration and willingness to adapt should be promoted when working with clients suffering from TS, to help them overcome psychosocial challenges faced on an everyday basis.

Similarly, for nurses who care for clients with TS, it is important to understand the client's family support system. Family is one of the key components responsible for promoting and maintaining the integrity of a client's health and the overall well-being of an individual. Nurses should recognize the importance of family in the client's life and provide appropriate care that defines the client's family dynamic. For example, family members may feel very confused, frightened, and uncertain when their child is first diagnosed with TS. There might be denial, anger, frustration, and feelings of guilt for the consequences that their child is suffering. Family members may be overwhelmed with feelings of uncertainty about what to expect with the child's health condition. This circumstance offers nurses opportunities to educate the family about TS. Nurses can educate parents about studies that suggest that TS is inherited via autosomal dominant transmission. Nurses can recommend parents for genetic counseling if needed (Castiglia, 1997). However, it is very important and helpful to reassure parents that children living with TS can make developmental progress and anticipate conventional life expectancies, developmental benchmarks, and career opportunities, and not mistreat for the client's medical condition (Sachapiro, 2002). It is also important to educate the client and their family members about the disease process. Educating parents about TS and helping them to identify signs/symptoms as well as duration and frequency of tics can prepare parents to face the challenges, consequences and uncertainty of the illness of their children with less anxiety and fear (Golder, 2010).

Appropriate education is also necessary for the client with TS. Racker (1997) indicates that this includes providing written "plans to provide access for clients with TS to educational programs/ protection from discrimination (Section 504) and (b) special education services in the educational setting" (as cited in Schapiro, 2002, p. 249). Similarly, Carter (1999) indicates that these protection acts provide services to meet the special needs of clients with TS, to help them succeed in their academic goals. The requirement for these written plans has brought several significant changes to the education system and accommodations offered to students with TS. Some of the exceptional changes include choice of seating in the class, parent feedback in choosing instructors for mentoring students with TS, authorizing students with TS to leave the classroom momentarily when they experience tic-like symptoms, supplementing with additional time on tests, and adapting homework according to a child's ability. Nurses play the role of advocate to support the client and their family members in providing appropriate tools for the overall success of the client and also assist in the decision-making process (Schapiro, 2002).

One of the important responsibilities of nurses is to address the clients' need individually, so that clients are not judged upon their medical condition. Though they may experience deficiencies in reading, writing, and arithmetic, clients with TS have IQs in the average range. (Castiglia, 1997). As the tics can range from mild to moderate and every client experiences them differently, these issues should be individually assessed and addressed. Likewise, to help cope with the stress, stigma and challenges

brought by TS to the lives of other family members, it is also important for nurses to assess family members' well-being. Nurses can find support group and social activities that can provide therapeutic and holistic care for family members' mental, emotional and physical well-being (Schapiro, 2002).

In this way, recognizing the importance of family, nurses should provide mental, moral and emotional support to the client, express sensitivity through active listening, inform the client and his/her family about the client's illness, show empathy, and nurture kindness throughout the care procedures. This care process will provide nurses with opportunities to explore, understand and respect the client's family values and culture and provide competent nursing care in the care setting.

Conclusion

Though Tourette Syndrome (TS), a chronic neuropsychiatric disorder, was first identified in 1825, the etiology is still not fully understood (Gelbart, 1998). Although there are genetic vulnerabilities, environmental factors may exacerbate the disease processes.

Nurses who work with children and teens with TS should be very understanding of the struggles and challenges the children with TS experience on both social and academic levels on an everyday basis (Ohm, 2006). Many of the symptoms of TS can be reduced by pharmacological intervention; however, it cannot be fully cured. All the aforementioned drugs used for the treatment of TS have severe side effects and drug-drug interaction, which causes some clients to become non-compliant with a medication regimen. Thus, the primary care provider often encourages and recommends that the client with TS seek non-pharmacological treatments as well (Schapiro, 2002).

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