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Striving to Maintain Normalcy Living with Addison's Disease

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Striving to Maintain Normalcy
Living with Addison’s Disease

Donita Mariegard

Carroll College
SIGNATURE PAGE

This thesis for honors recognition has been approved for the Department of Nursing.

Director

Date

Reader

Date

Reader

Date
Abstract

Purpose: Studies show a correlation between premature mortality rates in individuals diagnosed with Addison’s disease. Individuals with this diagnosis go from living active, busy lives to having difficulty maintaining work schedules and struggling to maintain normalcy in everyday life. The purpose of this study was to gain understanding of the issues and problems of living with Addison’s disease.

Demographics: Four adult individuals, one male and three females, one which resides in Australia, one in Canada, and the other two within the United States, participated in this qualitative research study.

Type of Study: Grounded theory qualitative research methodology was utilized.

Data Collection and Analysis: Data collection included in-depth personal interviews and correspondence, field notes, and theoretical memos. Classic grounded theory was used for data analysis.

Findings: The core category of this research was “Striving to Maintain Normalcy” with the following supporting categories: a) Finding a Diagnosis, b) Tolerating Fatigue, c) Experiencing Frustration, Fear, and Depression, and d) Coping with Finances, Workplace Environment, Spiritual Matters, and Support.

Nursing Implications: The findings of this study will increase the nurse’s understanding of Addison’s disease and bring awareness of the concerns and problems that individuals have living with this disease.
Generalization: The findings of this research cannot be generalized to all individuals diagnosed with Addison’s disease; however, more research will contribute to a better representation of results.
Acknowledgements

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Dedication

To my brother, Steve, who passed away October 1, 2009 of adrenal insufficiency. I love and miss you, and look forward to seeing you again someday.
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CHAPTER I
BACKGROUND

The diagnosis of Addison’s disease carries a significant risk of premature mortality (Bergthorsdottir, Leonsson-Zachrisson, Oden, & Johannsson, 2006). Addison’s disease is a disorder of the adrenal glands. These glands are located above the kidneys and are responsible for producing a variety of hormones, each producing many complex effects on the body (Berry, 2009). The incidence of Addison’s disease, or primary adrenal insufficiency, is approximately 11 in 100,000. Etiology of Addison’s disease varies. Most individuals are diagnosed with Addison’s disease between ages 30 and 40, and some research has shown Addison’s disease to occur three times more often in women than in men (Berry, 2009). During the last century, tuberculosis was a significant cause of the disease; however, most recent cases of Addison’s disease in developed countries are a result of autoimmune disease (Bergthorsdottir et al., 2006).

Addison’s Disease

Adrenal insufficiency can be categorized as primary or secondary adrenal insufficiency. Primary insufficiency occurs when the adrenal glands are destroyed, and the hormone amounts being distributed in the body are significantly lowered (Berry, 2009). Secondary adrenal insufficiency is more common and is caused by insufficient secretion of adrenocorticotropic hormone (ACTH) due to problems related to the hypothalamic-pituitary-adrenal axis (Whiteman, 2009). Clients with primary or secondary adrenal insufficiency require exogenous steroid replacement. Typical treatment includes intravenous hydrocortisone or oral prednisone to simulate
diurnal adrenal rhythm (Whiteman, 2009). Individuals who have been diagnosed with adrenal insufficiency (AI) currently achieve survival; however, treatment regimens may not aid in normal life expectancy or normal quality of life (Reisch & Arlt, 2009). Research indicates increased mortality in individuals with both primary and secondary AI, excluding comorbidities. Research also indicates reduced capacity for full-time employment as well as an increased rate of disablement pensions in those with AI (Reisch et al., 2009).

**Signs and Symptoms**

Signs and symptoms of Addison’s disease include weakness, hypotension, sinus tachycardia, fever, altered mental status, diarrhea, loss of body hair, myalgia, and in the acute stage, nausea and vomiting and abdominal pain (Thomson Micromedex, 2009). According to another source, symptoms of AI begin gradually and commonly start with chronic, worsening fatigue, muscle weakness, loss of appetite and weight loss. Symptoms can progress to low blood pressure which lowers upon standing, which can contribute to dizziness and/or fainting, irritability and depression, cravings for salty foods, hypoglycemia, and headache (Volfson, 2006). Since the symptoms of adrenal crisis can be vague, and may include sudden, penetrating lower back pain, severe vomiting and diarrhea, dehydration, low blood pressure or loss of consciousness, there is the potential for ignoring the signs/symptoms which could result in an addisonian crisis, or acute adrenal insufficiency, for which rapid medical treatment must be initiated or the result may be fatal (Volfson, 2006).
Suzuki et al. (2007) stated that numerous authors found that clients diagnosed with Addison’s disease commonly suffer from depression (Engel & Margolin, 1942; Cleghorn, 1951; Craddock & Zeller, 1952; Cumming & Kort, 1956; Kaushik & Sharma, 2003; Leigh & Kramer; 1984; Fava et al., 1987; Thomsen et al., 2006; Thomsen, Kvist, Andersen, & Kessing, 2006). In many cases, clients living with Addison’s disease experience psychiatric symptoms such as agitation, stupor, and refusal of medical treatment. Individuals with Addison’s disease may also present with psychiatric symptoms since a review of literature shows that disturbances in mood, motivation, and behavior may be associated with the disease (Anglin, Rosebush, & Mazurek, 2006).

Symptoms of Addison’s disease, whether physiological or psychological, can potentially develop into a life-threatening crisis (Suzuki et al., 2007). These issues could dramatically impact family dynamics depending upon the coping ability of the individual with Addison’s disease. Because of the potential for depression, further consults with mental health professionals may be appropriate.

**Impact on families**

Financially, the diagnosis of Addison’s disease can carry with it financial hardship. Diagnosing adrenal insufficiency may include laboratory tests as well as imaging exams (Berry, 2009). Also, because the etiology of Addison’s can vary from destruction of the adrenal glands to problems with the hypothalamus or pituitary, there may be necessary extensive testing/surgery. Specific tests that may be used to diagnose Addison’s disease are chemical stimulation tests; EEGs show brain activity; CT scans show the adrenal glands as well as muscles, joints, and bones.
Magnetic Resonance Imaging (MRIs) is also used to assess the adrenal glands (Micromedex, 2009). Individuals with minimal or no medical insurance coverage can have serious effects on finances, especially if the diagnosis involves surgery. AI symptoms can be vague and mask other physiological problems; therefore, diagnosis may be delayed, causing additional testing to determine the cause of the symptoms.

Impact on the families of clients with Addison’s disease may be more than financial. Spouses of individuals with Addison’s disease rated the consequences of the disease more seriously than their partners, (Heijmans, deRidder, & Bensing, 1999).

**Education**

When an individual has been diagnosed with AI, there is need for extensive teaching for both the person diagnosed with AI as well as the individual’s family/significant others. Teaching should include information concerning everyday medication dosing and route, how to adjust medication dosing for sick days, and how to assess for signs and symptoms of adrenal crisis. In the case of adrenal crisis, a rapid infusion of 5% dextrose in 0.9% sodium chloride should be started immediately to correct dehydration, hypotension, hyponatremia, and hypoglycemia (Micromedex, 2009). Teaching for family/significant others may also include potential psychological issues that can accompany AI. There may be a need for readjustment to living with someone with AI. Careful clinical assessment and client/family teaching are crucial to the health and well-being of the client.
CHAPTER II

REVIEW OF THE LITERATURE

The following is a review of literature that identifies the importance of client education and teaching, etiology of adrenal insufficiency (AI), the need for lifestyle modifications, psychological issues concerning AI and how this can affect quality of life, and symptoms and management of adrenal crisis.

Mortality

There is a higher incidence of premature mortality in patients who have been diagnosed with Addison’s disease (Bergthorsdottir et al., 2006). One study followed 1,675 patients with Addison’s disease between the years of 1987 and 2001. The National Hospital and Cause of Death Registers and the Swedish National Board of Health and Welfare were used to identify and track participants.

The average follow-up period was 6.5 years: 3.4 years for those patients who died and 7.9 years for those patients who survived the observation period. The study concluded the largest number of deaths were from cardiovascular (ischemic heart disease), followed respectively by malignant, endocrine, respiratory, and infectious diseases. The actual number of deaths was significantly higher than the number of expected deaths during this time period (507 actual deaths compared with 199 expected deaths). This study validated the thought that premature death was associated with Addison’s disease (Bergthorsdottir et al., 2006). Implications of this study indicate the need for extensive teaching, monitoring, and follow-up in individuals with Addison’s. There is a need to implement these findings into nursing practice since healthcare professionals have direct contact with these individuals and
must understand the importance of communicating these findings to those who need to understand their condition and know how to effectively and safely care for themselves.

**Diagnostic testing**

Secondary adrenal insufficiency relates to either a malfunction in hypothalamus or the pituitary glands. Since diagnosing secondary adrenal insufficiency is critical for survival, it is important to have the tools for diagnosing hypothalamic-pituitary-adrenal insufficiency (HPAI).

The purpose of the study was to compare the performances of standard-dose corticotrophin stimulation test (SDCT) and low-dose corticotrophin stimulation test (LDCT) when diagnosing HPAI. Based on the study’s findings, the LDCT was a better diagnostic tool in diagnosing HPAI (Kazauskaite et al., 2008). The methodology of the study was to search the PubMed database from the years 1966 through 2006 searching for articles with the keywords “adrenal insufficiency.” Studies with at least 10 subjects that were suspected to have HPAI were selected. The researchers then attempted to make contact with the principal investigator of each relevant study and requested information regarding the results. The implications of this study are important since there is an increased rate of premature death related to adrenal insufficiency; if patients can be tested for secondary adrenal insufficiency and treated, the rate of premature deaths may be lowered. The study’s findings for evaluating patients with possible HPAI suggest a three-step approach. The first is measuring a morning basal cortisol level; if this test is inconclusive, the second step would be to perform an LDCT. If this test is indeterminate, the study suggests an
insulin hypoglycemia test. The combination of these tests will accurately diagnose the majority of patients (Kazlauskaite et al., 2008).

Individuals who present with polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes syndrome (POEMS) have a high prevalence of endocrinopathies and need a thorough endocrine investigation (Gandhi, Basu, Dispenzieri Basu, Montori, & Brennan, 2007). POEMS syndrome has two major criteria and 1 minor criterion; the two major criteria include polyneuropathy and clonal plasma proliferative disorder, and the minor criteria include osteosclerotic bone lesions; Castleman disease; papilledema; organomegaly, including lymphadenopathy; edema, pleural effusion, or ascites; endocrinopathy; and skin changes, (Gandhi et al., 2007).

Through the Mayo Clinic dysproteinemia database, 170 individuals identified as having POEMS syndrome were selected for this quantitative study. From this group of participants, the study followed individuals from January 1, 1960 through June 30, 2006; however, because those evaluated after the year 2000 had a more extensive endocrine evaluation, the study focused on the 64 individuals treated in 2000 or later. Of the 64 participants treated in 2000 or later, 54 had a recognized endocrinopathy with the most common being hypogonadism. Other endocrine abnormalities found in the individuals with POEMS syndrome included thyroid, glucose metabolism, adrenal insufficiency, and calcium regulation. Multiple endocrinopathies occurred in 54% of the individuals. AI, which is defined in this study as an abnormal response of cortisol to stimulation with standard high-dose
synthetic adrenocorticotropic hormone (ACTH), was noted in 6 of the 9 (67%) participants tested (Gandhi et al., 2007).

This study is important because it confirms that endocrinopathy is the central aspect of this study and adrenal insufficiency, which previously has been infrequently reported in association with POEMS, may be more common than previously thought (Gandhi et al., 2007).

**Life style modifications**

The diagnosis of Addison’s disease requires treatment and life-style changes in order to achieve survival. Therefore, individuals with this diagnosis must make necessary changes to accommodate for the lack of glucocorticoids being secreted by the adrenal glands. These changes include: daily medication doses of glucocorticoid replacements, assessing themselves for signs and symptoms of adrenal crisis, carrying medical alert identification, and also carrying an injectable form of glucocorticoid replacement in case of crisis.

The diagnosis of Addison’s disease carries with it long-term life-style changes and adaptations for patients and family members (Hahner et al., 2007). This study indicated impaired subjective health status in those with Addison’s. The general self-perception in individuals with adrenal insufficiency was significantly impaired as evidenced by restrictions in activities of daily living and a high incidence of occupational changes that were associated with adrenal insufficiency (Hahner et al., 2007). This cross-sectional study followed 256 individuals with adrenal insufficiency. These individuals were contacted via mail and asked to complete three validated self-assessment questionnaires. The results were compared with age and
gender-matched controls drawn from the questionnaire-specific reference cohorts. Major findings indicated a significantly reduced subjective health status in those with Addison’s disease (Hahner et al., 2007). Implications for this study are relevant to health professionals since being aware of signs and symptoms of Addison’s disease could potentially save lives. Individuals with this diagnosis need extensive support and teaching regarding the diagnosis and all that it entails. It is important for individuals to understand correct medication dosing and route, on well days and sick days, and they must be able to assess for signs and symptoms of adrenal crisis. It is also important to wear medical emergency wristbands and carry an injectable form of steroid, in case of adrenal crisis. Since this study indicates those with adrenal insufficiency feel their overall health status is impaired, there is need for further teaching regarding psychological issues as well as physiological issues (Hahner et al., 2007). Referrals to licensed mental health professionals may be appropriate in order to maintain the mental well-being of the individuals. The study’s findings are helpful in assisting nurses to provide appropriate tools, resources, or teaching for individuals adapting to the life changes caused by Addison’s disease.

**Patient Education**

Another study suggested while adrenal crisis may be predictable, it is often undermanaged (White & Arlt, 2010). The findings of this study indicated patient education is vital for survival of individuals with Addison’s disease. Two separate postal surveys in 2003 and in 2006 were mailed to UK Addison’s disease Self-help group members regarding their experiences of adrenal crisis. The respondents were asked to identify all of the factors that led up to the adrenal crisis and subsequent
emergency response. In more than half of the reported adrenal crises, vomiting and/or diarrhea were responsible. The next most important risk factors reported by individuals were flu-like illness and associated major infections, followed by surgical procedures carried out without sufficient use of steroid cover (White & Arlt, 2010). Implications for clinical practice indicate patient teaching is crucial to inform Addison’s sufferers to report and receive treatment at the first sign of vomiting/diarrhea or flu-like symptoms. Also important during surgical procedures is the cover of steroid therapy. All health care professionals involved in the care of these individuals must be aware of the importance of steroidal coverage during surgery.

**Septic shock**

Individuals presenting with septic shock should be evaluated/tested for AI since there is a correlation between septic shock and AI. In recent years AI has been the focus of many studies and has been recognized in as many as 40% of those with septic shock. This is a particularly important finding since treatment of AI in individuals with septic shock has been associated with decreased mortality rates (Guillamondegui et al., 2009).

After IRB approval, the Vanderbilt University Level 1 trauma center database was reviewed for individuals admitted between the fiscal year 2002 through 2004. The following were criteria for inclusion in the study: greater than age 18; admission to ICU, length of stay greater than 24 hours but less than 7 days; ventilator requirements longer than 24 hours with ongoing pressor requirements; and participants who underwent cosyntropin stimulation test (CST). The CST was
completed based on clinical suspicion of adrenal insufficiency on individuals on mechanical ventilation and those with systemic inflammatory response syndrome with vasopressor dependence or unexplained hypotension which required ongoing resuscitation after 24 hours in the ICU. The CST was obtained only if the individual had completed the initial resuscitation process, but was still showing signs of hypotension and shock (Guillamondegui et al., 2009).

During the time described, 7,949 individuals were admitted with 1,795 intubated patients requiring ICU placement. Of these, 137 trauma patients had CSTs performed. This group was divided into two groups based on whether or not they suffered from AI. Eighty-two of these individuals had AI. Sixty-six patients having AI were treated with hydrocortisone based on the practice management guideline, and sixteen of the patients with AI were not treated with hydrocortisone therapy. The untreated patients had significantly higher mortality (Guillamondegui et al., 2009).

The treatment of AI in these individuals was based on published reports and consisted of 50 mg hydrocortisone IV every 6 hours for one week or until weaned from the hydrocortisone (if earlier than one week). Steroid therapy has been shown in numerous studies to decrease time to vasopressor withdrawal in the setting of septic shock. This is an important study since early diagnosis and treatment of AI may lead to decreases in the mortality rate of this patient population (Guillamondegui et al., 2009).

One study concluded there was a high incidence of adrenal insufficiency with Surgical Intensive Care Unit (SICU) individuals that were greater in age than 55 with post-operative hypotension requiring vasopressors (Rivers et al., 2000). While AI is
rare in the general population, up to 28% of critically ill individuals often are found to have unrecognized AI (Rivers et al., 2000). To be enrolled in this study, individuals had to be postoperative patients greater than age 55 who experienced hypotension requiring vasopressor therapy subsequent to adequate volume resuscitation within 24 hours of SICU admission. Excluded from this study were individuals positive for HIV, individuals with known preexisting adrenal disease or adrenalectomy, administration of hypnotic drugs prior to general anesthesia, administration of steroids during surgery, or having taken steroids within the three months prior to admission. There were 104 individuals participating in the study with a mean age of 65.2, plus or minus 16.9 years. Of the 104 group participating, adrenal function was abnormal in 34 of the individuals. Screening for AI in all ICU individuals may not be an appropriate measure; however, since age has been shown to increase risk of AI, those over the age of 55 and who exhibit signs of hypotension after adequate volume resuscitation should be considered at high risk for development of AI (Rivers et al., 2000). SICU nurses could impact the lives of individuals by being aware of these factors and suggesting diagnostic measures related to AI in these types of individuals. Survival of individuals with adrenal insufficiency is routinely achieved; however, this does not necessarily mean a normal quality of life or normal life expectancy.

**Quality of Life**

Quality of life has shown to be impaired as evidenced by an increased rate of disablement pensions and reduced capacity for full-time employment (Reisch & Arlt, 2009). Treatment for adrenal insufficiency currently involves hydrocortisone, (ie.,
cortisol) and as a rule is chosen for replacement therapy. For primary insufficiency, a daily dose of hydrocortisone 20-25mg is recommended, and for secondary insufficiency a daily dose of 15-20-mg is recommended (Reisch & Arlt, 2009). It is important to consider other medications being taken when considering the glucocorticoid dose. Most often, the initiation of glucocorticoid replacement in those newly diagnosed with adrenal insufficiency due to hypopituitarism should precede the start of L-thyroxine replacement (Reisch & Arlt, 2009). Careful clinical judgment and assessment are important in the monitoring of those with adrenal insufficiency since there are no reliable markers of glucocorticoid action. Symptoms of under-replacement include fatigue, nausea, myalgia, and weight loss while symptoms of over-replacement include weight gain, central obesity osteoporosis, impaired glucose intolerance, and hypertension (Reisch & Arlt, 2009). Health care professionals must ask detailed questions regarding the individual’s past history as well as current signs and symptoms in order to avoid adrenal crisis. Health care providers should also ask about the individual’s ability to cope with stress and whether the individual under- or over-managed glucocorticoid replacement by self-adjusting doses. At every visit, teaching is critical for the individual as well as family members so adrenal crisis may be avoided (Reisch & Arlt, 2009).

**Adrenal Crisis**

Adrenal crisis is a life-threatening condition that can be brought on by stress during surgery in those with AI (Yong, Marik, Esposito, & Coulthard, 2010). This study explored the possibility of preventing adrenal crisis in those with adrenal insufficiency by administering high doses of steroids prior to surgery. The selection
criteria were randomized, controlled trials that compared supplemental perioperative administration of steroids with administration of a placebo in adult participants taking maintenance doses of steroid therapy prior to surgery. There were two trials involving 37 participants; however, the results of this study were inconclusive. There was no evidence showing that the use of perioperative steroids in patients with AI decreased risk of adrenal crisis. This could be due in part to the small number of participants (Yong et al., 2010).

**Implications for Nursing Practice**

Though the prevalence of Addison’s is disease relatively low, and the symptoms of Addison’s disease can be extremely vague, it is vital for the nurse to understand the pathology of the disease, recognize and assess initial signs and symptoms, and alert physicians to perhaps complete diagnostic testing for Addison’s disease in individuals presenting with symptoms such as fatigue, nausea, vomiting, headache, weakness, hypotension, sinus tachycardia, fever, altered mental status, diarrhea, loss of body hair, and muscle weakness.

**Summary**

The effects of Addison’s disease on individuals are extensive. It is important for nurses to recognize the signs and symptoms so that early detection, diagnosis, and treatment are possible. It is also important for nurses to understand the importance of teaching for individuals with this diagnosis since there are many variables such as sickness which can cause the onset of adrenal crisis, at which time, the individual’s dose and route of medication administration must be altered to avoid crisis and potentially death. After diagnosis, extensive teaching is required in order
to enhance the individual’s quality of life. Research with regard to individuals’ lived experience with Addison’s disease was not included in this review of literature.
CHAPTER III

Methodology

Grounded theory is the discovery of theory from data (Glaser, 1967). Individuals with Addison’s Disease and their families/significant others, face additional challenges and obstacles than those without the diagnosis. It is important to study Addison’s disease and its effects on individuals because although it is not widely prevalent, it has life-changing and potentially life-threatening effects on individuals.

Purpose

The purpose of this study was to gain understanding of the issues and problems of living with Addison’s disease. Since the diagnosis of Addison’s disease carries a significant risk of premature mortality (Bergthorsdottir, Leonsson-Zachrisson, Oden & Johannsson, 2006), it is important for nurses to understand the issues which affect individuals that have been diagnosed with Addison’s disease. There are many potentially life-changing/threatening elements of this disease, and the diagnosis requires coping and compliance for survival. The researcher’s purpose was to gain understanding of how those with the disease perceived the physical, emotional, spiritual, and financial impact of the disease. Pathology of this disease primarily results from insufficient levels of glucocorticoids, mineralcorticoids, and androgens, and in order to survive, individuals must take daily oral hormone replacement therapy. Those diagnosed with Addison’s disease will have significantly altered lifestyles and depending on each one’s coping ability, will adapt and make necessary changes, or will face adrenal crisis and potentially death.
Characteristics of Participants

Criteria for being interviewed were English-speaking individuals over the age of 18, individuals of both genders that had been diagnosed with Addison’s disease, and demonstrated a willingness to participate in the interview. The researcher recruited volunteers in the Northwest area of the United States, Canada, and Australia by word of mouth as well inviting participants through a support group network. The researcher also contacted physicians who specialized in endocrine disorders in order to recruit participants with Addison’s disease. One contact was made through a local endocrinologist; the other contacts were made through a support group network. The researcher posted a notice of inquiry on the support group network and potential participants contacted the researcher. The researcher obtained voluntary informed consent in a manner consistent with the requirements of the Institutional Review Board (IRB) before beginning the study.

Data Collection

Prior to data collection, IRB approval and informed consent were obtained. The collection of data was done primarily through interviews with people who had this disease or people who lived with someone diagnosed with this disease as well as interviews with physicians that specialized in endocrinology. Qualitative research was used to structure knowledge since the crucial elements of sociological theory often are best found using a qualitative method (Glaser, 1967). The interview process was the most important component in gaining understanding of the disease from the individual’s point of view. Three of the participants were recruited through a computer support group network. The researcher posted a notice on the support
group network regarding the study on Addison’s disease, and participants were requested to respond via email if interested in participating in the study. The researcher then emailed the informed consent and a list of questions about Addison’s disease. Before answering the questions, the participants were asked to sign the informed consent and email it to the researcher. One interview was completed in person using observation, and open- and closed-ended questions. The interview was audio-taped and transcribed for the purposes of coding and analysis of data.

**Data Analysis**

Grounded theory was used in the collection of information for research. According to Glaser, “…grounded theory is the systematic generation of theory from data acquired by a rigorous research method” (Glaser, p. 3). This was the gathering of data from the individuals with the diagnosis of Addison’s disease. A coding process was used to identify common themes among the participants’ reflections.

**Ethical Considerations and Confidentiality**

Before initiating the research project, the researcher completed the NIH sponsored "Human Participants Protection Education for Research Teams" training course. The Carroll College Institutional Review Board approval was obtained prior to the initiation of the study. Confidentiality was maintained by keeping client-identifiable information in a locked cabinet and password-protected computer. Once the study was concluded, the data collected from individuals were destroyed.
Limitations

The study was limited by the small number of individuals interviewed for this honors thesis research project. The views of those participating in the study do not necessarily reflect the opinion of the majority of people living with Addison’s disease.
CHAPTER IV
Research Findings

The purpose of this research was to gain understanding of the issues and problems of individuals living with Addison’s disease using qualitative grounded theory methodology. The core category of this research was “Striving to Maintain Normalcy” with the following supporting categories: a) Finding a Diagnosis, b) Tolerating Fatigue, c) Experiencing Frustration, Fear, and Depression, and d) Coping with Finances, Workplace, Spiritual Matters and Support. These categories will be further discussed, detailed and validated by individual quotes and paraphrases of the research participants.

Core Category: Striving to Maintain Normalcy

Participants described their lives prior to being diagnosed with Addison’s disease as being successfully employed, self-sufficient, and independent. Post-diagnosis they described themselves as being unable to work consistently or make commitments to social activities due to unpredictable symptoms and management. All participants strived to maintain normalcy in their lives. This core category was seen throughout all the supporting categories: a) Finding a Diagnosis, b) Learning to Tolerate Fatigue, c) Experiencing Frustration, Fear, and Depression, and d) Coping with Finances, Workplace and Spiritual Matters. Every day participants strived to maintain normalcy in their lives. One participant identified lifestyle changes due to the disease: “I don’t commit to and social activities, because I may be having a bad day. I don’t go partying and dancing like I used to. I haven’t returned to work as of yet.” Another participant described feeling vulnerable living with a disease that was
potentially deadly if mismanaged and stated, “There’s certainly more fear…and it’s scary.” Participants with Addison’s disease must self-regulate with appropriate steroid dosing daily and adjust according to levels of stress or illness. This takes being aware of signs and symptoms of stress and illness. The presenting signs and symptoms, which are potentially life-threatening, can be vague. Participants living with Addison’s disease have many commonalities that range from receiving the initial diagnosis to every day coping with the disease. Participants with this disease appeared to struggle as it related to achieving physical and psychological stability. For the participants, the diagnosis of Addison’s disease dramatically changed their lives from living active busy lives, to being dependent, vulnerable, and struggling to maintain everyday life without going into adrenal crisis. One participant described the complexities of day-to-day management of Addison’s disease in the following way (quotes from the participant were emailed directly to the researcher so grammar and punctuation are exactly per participant):

My day-to-day management revolves around a dynamic control model. I have developed this myself after quite a few years of getting it wrong. I regulate cortisol intake normally as 3 doses of Hydrocortisone [HC] per day. Depending on the physical severity of the day I pre-plan to take 35 or 45mg in declining dos[e] sizes. I also buffer these doses with food to push the diurnal curve into a shape that best meets that of a normal human. I also have to regulate my water levels. HC contains a mineralocorticoid and since I “reset” every night, my morning HC dose elevates the correct level. If I drink too much, I bloat and go very high blood pressure [BP]. As a general rule this
means I limit water intake up until about 10AM and then drink to capacity. In some cases I need to eat salt in the afternoons as it goes the other direction. Handling stress cortisol needs I do a little different from most. Instead of elevating HC I take Prednisolone in addition to it, in a ramping down style. There is some trial and error in this, but I rarely more than triple my cortisol levels for more than a few days. I have also found that I can't simply respond to fever as I need enough cortisol to even cause an immune reaction. If I suspect one I may take 12.5mg of Prednisolone and see if I immune react, then dose according to that reaction. (Prednisolone comes in 25mg pills here).

It is interesting to note that although untreated primary results in water volume loss, treated secondary can result in the same problem if you use a low mineralocorticoid content replacement steroid like Prednisolone or Dexamethasone. This would appear to suppress what ACTH production you have to even lower levels and thus aldosterone drops off. I know some secondary people on Prednisolone that have to salt up like I do. I have a BP machine and I should also get a glucometer. Adrenal based fatigue is often seen in low levels as these. I also use an anti-histamine 2-3 times per week so I can limit the need for the anti-inflammatory effect of the HC. (Since pills create cyclic blood levels) I have a permanent sore throat. By the way I arrived at my HC dosing based in reading lots of medical papers and slowly bringing my exercise rate up. I still however am not stable enough for a full work week. In worst case scenarios I have a set of trigger events that I “listen to” as far as the need for extra dosing is needed. I never trust medical advice
as it always seems flawed. I will however get to a hospital if I can't seem to control it myself. My main triggers are a decrease in eye movement speed (eye drag), unable to get warm or highly fluctuating body temp, very sleepy, low BP, brain stupidity and limb coordination failures.

This same participant also stated:

I have found that since it is fairly rare, knowledge even in the specialist community is rarely based on experience. It would seem that the doctor can certainly keep you alive but quality of life seems to be badly understood and managed.

Another participant stated: “I never leave home without some extra prednisone in my purse. I try to avoid stressful situations.”

Several of the participants voiced thoughts and feelings of fear resulting from the lack of knowledge among healthcare practitioners. Frustration resulted from the inconsistencies among healthcare practitioners regarding diagnosis, treatment, and prognosis. One participant stated: “It’s so rare and uncommon, that most doctors are not very familiar with it, endocrinologists as well. My family doctor still asks me at every visit, how long am I going to be on the prednisone for…‘forever.’”

Another participant stated:

It is very frustrating how little doctors and nurses actually know about Addison’s and how we are treated. I hear horror stories all the time on my forum as to how many of us are treated because there are so many unknowns and not enough data out there.
Supporting Category: Finding a Diagnosis

Since signs and symptoms of Addison’s disease are vague and can range from nausea, vomiting, and fatigue to mood disturbances, individuals with the disease often are delayed in receiving a diagnosis and the starting of treatment. Participants voiced differing thoughts on how they were diagnosed with Addison’s disease.

One participant discussed the length of time from start of symptoms to diagnosis, stating: (grammar and punctuation as written by participant), “About 6 months before a positive diagnosis though I was “crashing” maybe 3 afternoons in 5 with extreme tiredness, dehydration and strange heart feelings.” One participant stated:

I was diagnosed in 2008, I was at deaths door, I had been sent home from a local hospital, they knew that I had severe gastroparesis (paralyzing of gastric system) and I couldn’t even keep water down, but they didn’t know what else to do with me so they basically sent me home to die. My husband wouldn’t give up and took me to a bigger hospital about an hour away from where we live. They have a wonderful endocrinology group that figured out within about 72 hours that I had Addison’s disease and that I was very nearly dead. Found out the pharmacy had filled the dosage wrong and I was taking a 10th of what I was supposed to be taking and I had gone into crisis, still couldn’t feel better ended up with shingles, thrush and in the hospital in another crisis in there for about 3 ½ weeks, they had to put in a j-tube and while they were in there found 2 hernias and scar tissue they had to clean up.
Unexplained fatigue is a common symptom of individuals with Addison’s disease; however, since this symptom is vague, it is sometimes overlooked as an indication of the disease and therefore is not correctly diagnosed in a timely fashion. In one case, the participant asked her endocrinologist to check her cortisol levels, and it was determined she had almost no cortisol in her system.

One participant described the following:

I wish there were more research monies being put into it. Um, I think there absolutely has to be a test developed. It’s an easy automatic test because again if that many people die in an ER, and they don’t die [from] what they came in, then, ER docs need to be able to recognize it very quickly. Um, people need to know whether or not they have it very quickly, so they can take preventative measures.

Another participant received a diagnosis of secondary adrenal insufficiency as a result of pituitary macroadenoma resection surgery. Regarding her diagnosis, she stated the following:

It was a possible risk after my surgery. And, 6 days after my surgery, my endocrinologist called and told me I had it, and to start taking HC. I thought my symptoms of fatigue, no appetite, weight loss, were due to my surgery. I really had no idea it was AI.

Another participant’s thoughts regarding the diagnosis of Addison’s disease: “It’s a complicated diagnosis, and hard to explain. They know it’s why I’ve gained weight, and that my body can’t deal with stress, that I have to take medication daily for it, and that it could be life threatening.”
Frustration was compounded by extreme fatigue experienced by participants. For the participants involved, the diagnosis of Addison’s disease dramatically changed their lives from living active busy lives, to being dependent, vulnerable, and struggling to maintain everyday life without going into adrenal crisis.

**Supporting Category: Tolerating Fatigue**

Fatigue was one of the most common themes among participants. Often the fatigue was experienced prior to diagnosis and was one of the signs and symptoms that pointed to Addison’s disease. The fatigue was described by participants as “extreme tiredness,” “crashing,” “unpredictable fatigue patterns,” and having “less energy.” Even post-diagnosis, participants described fatigue as a routine part of life. Individuals with Addison’s disease described being fatigued or extremely tired. When asked about her support system, one participant stated: “They are very supportive though it is hard for them to understand what I go through with fatigue and stress dosing and having to carry an emergency injection with me all the time.” Another participant stated:

I was having unexplained fatigue, low immune repair rates, gastric upsets, and minor AI symptoms for years. About 6 months before a positive DX though I was crashing maybe 3 afternoons in 5 with extreme tiredness, dehydration, and strange heart feelings.

Another participant stated:

Well, in simple terms I haven't been able to work since January 2010. The problem is more one of unpredictable fatigue patterns…and then, this past year, I noticed that my, um, fatigue levels were going way up. Um, I have a
number of other autoimmune disorders, so I wasn’t sure whether that was, was causing it, or whether I needed to really work on stress reduction.

Another participant voiced the following with regard to living with fatigue:

Now, it doesn’t mean my system is the same, but because of the sort of long onset of the fatigue, and the, um, I’ve started having some memory lapses, um, just wasn’t functioning as well as I expected myself to or normally did…um, a couple of days when I’ve felt just unusually fatigued, sort of beyond reason, I’ve added another five milligrams, and they’ve kept me on a, at first I was on twenty milligrams in the morning, ten at night. Um, they’ve kept me now on a ten at- at um, midday as well.

Still another participant stated, “I don’t commit to any social activities, because I may be having a bad day. I don’t go partying and dancing like I used to. I haven’t returned to work as of yet.” One participant stated, “I was just overwhelmed by tiredness…I was laying down and trying to nap, which just isn’t me.”

Fatigue was a consistent characteristic among the participants living with Addison’s disease. At times fatigue was used as a diagnostic tool; other times it was the result of a dosing/mis-dosing issue, and at other times it was a result of adrenal crisis.

**Supporting Category: Experiencing Frustration and Fear and Depression**

The majority of the participants voiced a common theme of Addison’s disease being both frustrating and frightening. Participants described their lives pre-diagnosis as being employed, self-sufficient and independent, and post-diagnosis being unable to work, commit to social activities and feeling vulnerable. One
participant described the feeling: “Um, there’s certainly more fear…um, and it’s scary….”

Another participant stated:

Low levels cause a fatigued/depressed state and like many other AI people, my initial diagnosis included clinical depression….Pre-diagnosis I did feel pretty bad and my GP did suggest medicating for depression. Post-diagnosis I certainly felt depressed on odd occasions as dosage didn't quite match, but being aware that it was a chemical balance thing (i.e., logic/science based) made it easy to ignore.

With regard to the experiencing of frustration and fear, one participant stated:

Um, blood pressure was very low, um, my pulse was very high. Um, he had me stand up, blood pressure dropped even lower, pulse went up even higher, and um, he said, you’re in adrenal crisis; you need immediately, need to change your dosing….

Another participant stated: (grammar and punctuation as written by participant)

So I picked up my new prescription and proceeded to take it for the next two weeks and woke up one morning unable to breathe in excruciating pain (muscles and joints seized up), nausea, and vomiting. My husband rushed me to family doctor about 5 minutes away as they are aware and they gave me my emergency injection, however I just couldn’t get to feeling better. Found out the pharmacy had filled the dosage wrong and I was taking a 10th of what I was supposed to be taking and I had gone into crisis, still couldn’t feel better, ended up with shingles, thrush and in the hospital in another crisis in
there for about 3 ½ weeks, they had to put in a j-tube and while they were in there found 2 hernias and scar tissue they had to clean up. The crisis you go into is horrible, there is no describing how frightening it is.

One participant described one of the frustrating situations he experienced: “The first major “attack” was in fact on my honeymoon in 2004. The first two nights were actually spent in hospital.”

Another participant discussed her emotions concerning the frustration of Addison’s disease:

I guess the hardest parts for me is the frustration. Of not knowing, for sure what caused this, if it’s going to go away or if it’s not, uh, how to treat it on an everyday basis, what to do if I get sick, um, the uncertainty if I get injured. I now have a um, a pendant that I wear in the house, a lifeline type pendant. It’s a little larger because it’s one I can talk into, so that I can say, I’m in the garage and I can’t get up or, um, I’ve fallen and call my husband at this number, or, those are not things I expected to be doing at sixty-five. I thought I’d be doing that at eighty-five.

One participant stated: “It is a very frustratingly, lonely disease because it is so rare.”

Another participant voiced frustration about the lack of knowledge regarding this issue: “I think the most frustrating part for me is that I like to know things.”

The frustration and fatigue that accompanies Addison’s disease contributes to feelings of depression. One participant stated: “My depression level is higher….”

Another participant stated: “I became very depressed. I am on anti-depressants, and
have had counseling. I believe it’s part of the grieving process...of a healthy body.”

One participant voiced the following:

Dr. Christopher Corsi, and, um, he felt that most likely because I’ve had numerous steroid injections because of um, other autoimmune conditions, the arthritis, I have a, what’s referred to variously as HOAB 27 disease, or um, sometimes as um, seronegative inflammatory arthritis, um, right now, uh, referred to most often as ankylosing spondylosis, or spondiolitis, um, which is another autoimmune (thing) that affects organs but affects especially in women, joints and, and so forth so, and I’d had that since I was a child.

This same participant also voiced the following: “But certainly my anxiety is higher. And my feeling of being vulnerable and fragile is higher.”

Individuals with Addison’s disease not only face ongoing physiological stress; they also experience financial difficulties, issues in the workplace, and must be aware of the signs and symptoms that accompany adrenal crisis. Several of the participants verbalized feeling alone and vulnerable; one stated:

Because, according to um, what Dr. Corsi has told me, if I should have an accident, where I’m seriously injured, then I have fifteen minutes to get a shot. So I carry the vial and the needle with me, and the instructions. It’s to go into the thigh. And it if doesn’t happen during that time then I die. So that’s not really time to call 911 and get somebody there. Whoever finds me has to do it right then. And if I start if I get the flu, and the vomiting, get severe diarrhea, then I have to up the medication right away. So, that’s not my favorite. I like to be in control. Hey. It’s a little more difficult with this.
Supporting Category: Coping with Finances, Workplace, Spiritual Matters and Support.

Addison’s disease affects individuals in many ways. Not only is the diagnosis potentially life-threatening; it impacts the individual’s ability to function in a work environment, which can affect the individual’s ability to pay for services related to the diagnosis and treatment of Addison’s Disease. Concerning spiritual matters, participants had conflicting views. Some drew strength from faith in God, whereas one participant stated he was an atheist. He stated the following:

I am an atheist as such so the definitions of spiritually may be different. To clarify first though I think you are asking how my life “feels” coping with the disease and for that matter how my body reacts physically to emotional states….I have come to believe through my own experiences and learnt knowledge of CNS control of the hypothalamus that my brain can influences my entire endocrine system. At a physiological level for example I know that emotional stress response still produces more ACTH/Cortisol. In simplistic terms, thinking positive can make me feel better. Over the years I also developed brain based pain control techniques that still work.

Another participant stated: (grammar and punctuation as written by participant)

“No, it has not affected me spiritually. I’ve always had strong faith and never ask ‘why me?’ because really ‘why not me?’ And being a nurse, I was already aware of how precious life is, and good health.” Regarding her faith, another participant stated: “…my faith in God that all things good come to those who believe in God.”

Individuals generally had support systems in place in order to cope with the realities
of Addison’s disease. For one participant, her family was a source of support, although she also joined an online support group. She stated the following:

That is why I joined the support group online that I did, so I can talk to others. Addison’s is a very individual disease and each person reacts differently to different types of stressors and each person reacts differently to the steroids.

When each participant was asked if they knew of anyone else with the same diagnosis, their answers were similar. One participant stated: (grammar and punctuation as written by participant) “My sole contact with other AI people has been through online means, I have never met anyone in person that has it. When we do it will be party time!” One participant stated she had not yet found any support groups because her healthcare providers were not in complete agreement, and still trying to provide an accurate diagnosis.

Being diagnosed with Addison’s disease had an impact on the employment opportunities for participants. Prior to the diagnosis of Addison’s disease participants were able to maintain full-time employment, however, after being diagnosed, were either not able to work or very limited in employment opportunities. One participant stated (grammar and punctuation as written by participant):

Well in simple terms I haven't been able to work since January 2010…but to be honest I think there is something else affecting my AI that if resolved would allow a better chance at work. For the 4 years 2005-2009 I was in fact in full-time work without too many problems…Since Feb 2009 I have more or less lived off savings and a small amount of telecommute contract work.
Feb 2009 also saw my marriage breakdown so that also had a huge financial impact. In the last month I have actually applied to the Australian equivalent of SSA (Centrelink) for assistance in finding work that will fit my capabilities. This assistance is also financial... In Australia the health (Medicare) and employment social security systems are joined. There is emphasis on finding any work or even community service that can be fitted with your capabilities. I for example may be able to volunteer to teach elderly people computer skills or even work in a public library.

For individuals whose identity is associated with employment, Addison’s disease can be particularly devastating. Another participant who had previously been self-employed stated: “I’ve closed my office now....”

For some individuals, not being able to maintain employment is disrupting, emotionally and financially difficult; depression can follow and this can impact all other areas of individual’s lives. Support is an important aspect of living with Addison’s disease. Whether the support is of a spiritual nature or the individual receives support from friends, family, or employers, it is vital to individuals with Addison’s disease to receive appropriate support. Nurses also provide an important avenue of support and have the potential for assessing, influencing, and linking individuals with a helpful, supportive network.
CHAPTER V

Discussion

The purpose of this study was to increase knowledge and understanding of the experience of living with Addison’s disease. The lived experience of being diagnosed with Addison’s disease carries with it life-long pharmacological treatment and the need for training and education concerning signs and symptoms of adrenal crisis. The core category of this research was “Striving to Maintain Normalcy” with the following supporting categories: “Finding a Diagnosis,” “Tolerating Fatigue,” “Experiencing Frustration, Fear, and Depression,” and “Coping with Finances, Workplace Environment, Spiritual Matters and Support.”

Primary adrenal insufficiency is primarily caused by an autoimmune destruction of the adrenal cortex (Bensing et al., 2008). Research has shown that individuals with autoimmune adrenocortical insufficiency are known to experience reduced quality of life and reduced working ability (Bensing et al., 2008). One Swedish research study reported a more than two-fold increased mortality risk in individuals with primary adrenocortical insufficiency (Bensing et al., 2008).

Secondary adrenal insufficiency can be caused by an abnormal growth in the pituitary or hypothalamus glands, congenital deficiencies, and long term use of glucocorticoid medications such as prednisone can interfere with normal pituitary function and is the most common cause of secondary adrenal insufficiency (Nieman, 2011).
Nursing Implications

It is vital that nurses recognize signs and symptoms of Addison’s disease and are able to assess for adrenal insufficiency. Though symptoms may be vague such as fatigue, weight loss, anorexia, nausea, vomiting, diarrhea, mental status changes, abdominal pain, fever, and muscle weakness, the outcome can be fatal if the individual has Addison’s disease and does not receive appropriate treatment (Whiteman, 2010). It is also vital for nurses to recognize psychiatric symptoms in those with adrenal insufficiency. These symptoms may include 1) mild to moderate organic brain syndrome, which manifests as memory impairment that can progress to confusion, delirium, and stupor; 2) depression which may manifest as apathy, poverty of thought, and lack of initiative; and 3) psychosis which can manifest as social withdrawal, irritability, negativism, poor judgment agitation, and hallucination (Kaushik, 2003). The psychiatric symptoms may occur early in the disease and may in fact predate other physical findings which may contribute to the difficulty in diagnosing Addison’s disease (Kaushik, 2003).

For individuals, the greatest impact of a disease lies in the effect it has on the person’s ability to continue with a ‘normal’ daily life (Hale, Treharne & Kitas, 2007). Nurses not only need to understand and be able to assess for signs and symptoms of adrenal insufficiency in those not yet diagnosed; they also must be able to connect with the individual who has already been diagnosed with Addison’s disease and establish a trusting relationship in order to support the individual physiologically and psychologically. Because individuals with Addison’s disease can appear irritable and may present with signs and symptoms of psychiatric disorders,
the nurse must perform a careful assessment, encourage follow-up care, and assess for knowledge, and compliance with medication doses and route.

**Summary**

Addison’s disease is a serious, life-threatening disease. Individuals diagnosed with Addison’s disease and family members, require significant understanding, empathy, treatment, and teaching. Since Addison’s disease is often unrecognized in early stages, it can present as a life-threatening crisis (Ten, 2001). It is important to note also that individuals over the age of 60 seem to be at higher risk of adrenal crisis (Reisch & Arlt, 2009). There are significant adaptations required in order to manage the illness effectively and attain normalcy. According to one study, fatigue is the number one most important health-related quality of life issue, although symptoms varied with individuals (Lovas et al., 2010).

Addison’s disease very often has psychological effects on individuals. Physicians need to be aware that Addison’s disease may present with psychiatric symptoms only and maintain a high index of suspicion for this potentially fatal condition (Anglin et al., 2006). It is important for nurses to understand that Addison’s disease symptoms may present as either psychological or physiological. Participants voiced frustration with the seeming lack of knowledge on the part of physicians and endocrinologists to be able to accurately diagnose and treat this disease. This could be due in part to the vagueness of symptoms, and because some of the presenting symptoms like depression and muscle weakness could be attributed partly to fatigue that was a commonly expressed category with most of the participants.
References


