Diagnoses Associated with Intellectual and Developmental Disabilities in Adult Decedents:
A Secondary Analysis of Healthcare Cost and Utilization Project National Inpatient Sample (HCUP/NIS) Data

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## Table of Contents

- Abstract 3
- Dissertation Proposal 4
- Summary of Changes from Proposal 35
- Slide Presentation 36
- Dissemination Plan 51
Abstract

Purpose: To identify primary and secondary diagnoses preceding death among adults with and without IDD who died during hospitalization.

Specific Aims: 1) to describe the commonly reported base diagnostic related groups preceding death among decedents with and without IDD who died during hospitalization in 2019, 2) to determine which base-DRGs had a higher prevalence rate among adults with IDD than among adults without IDD, controlling for age, gender, race, urbanicity of person’s residence, US census division of hospital, and mean income of person’s zip code, and 3) to use the base-DRGs and ICD-10-CMs to examine the conditions of the Fatal Four/Five as conditions of concern preceding death in decedents with and without IDD.


Results: Identified fourteen primary diagnoses at the time of death for decedents with IDD that are represented at a higher percentage than for decedents without IDD and have a significant odds ratio for IDD diagnosis.

Conclusion: A new set of conditions is proposed to assist nurses in reducing preventable deaths in decedents with IDD. Dehydration, GI obstruction, respiratory infection, seizures, and sepsis, will be known as the IDD Concerning Conditions. Aspiration, constipation, and GERD, the IDD Contributing Conditions, are conditions that do not cause death in themselves but contribute to the development of at least one of the IDD Concerning Conditions, which do cause death.

Keywords: Intellectual disabilities, Mortality, IDD Concerning Conditions, IDD Contributing Conditions
Dissertation Proposal

Making up 1% of the population (Maulik et al., 2011), and with a prevalence that is increasing (Bilder et al., 2013; Cooper et al., 2004; Zablotsky et al., 2019), adults with intellectual and developmental disabilities (IDD) have poorer health outcomes (Ervin et al., 2014; Lauer & McCallion, 2015), bear a higher percentage of preventable diseases (Glover et al., 2017), tend to have more complex, multi-faceted medical conditions (Auberry, 2018), and suffer lower life expectancies (Arvio et al., 2016; Glover et al., 2017; Heslop et al., 2014; Landes et al., 2019; Lauer & McCallion, 2015; Maulik et al., 2011; O’Leary et al., 2018) than people who do not have IDD. In the past, discrepancies in life expectancies and overall health status of adults with IDD have been attributed to poor sanitary conditions, lack of adequate medical care, crowding, poor nutrition, and other aspects of living in an outdated institutionalized environment (Janicki et al., 1999). However, since the 1960s there has been a major shift towards encouraging adults with IDD to live in the community and pursue a more conventional lifestyle (Chowdhury & Benson, 2011). Today, community settings might include a group home, a family home, section-8 housing, an assisted living facility, or a skilled nursing facility. Although the change of residing in institutions to community living for adults with IDD was driven by the hope of better quality of life (Chowdhury & Benson, 2011), mortality has not improved with this move (Hayden, 1998; Lerman et al., 2003) and the healthcare system itself is partly to blame (National Council on Disability, 2009). The purpose of this secondary data analysis is to identify and describe common diagnoses and factors associated with having IDD among adults who died during hospitalization. Recognition of patterns of co-occurring diagnoses and other factors may prompt earlier recognition and intervention, furthering efforts to decrease the disparities in health outcomes reported for this vulnerable group.

A developmental disability is any disability that prevents a person from reaching a developmental milestone (National Institutes of Health, 2016). These conditions are often present at birth, and can include a physical disability, an intellectual disability, or an emotional disability (National Institutes of Health, 2016). An intellectual disability, a subset of the developmental disabilities, begins before the person turns 18 years old, and affects both a person’s intellectual functioning and adaptive behavior (National Institutes of Health, 2016). This intellectual disability affects the person’s ability to learn, reason, and problem solve, and affects their ability to apply their knowledge to manage everyday social and life skills (National Institutes of Health, 2016).
The heterogenous group of intellectual disabilities affects people in dramatically different ways depending on the etiology and severity of their disability, as well as the resources that the person has available to manage the sequelae (Coppus, 2013).

Most undergraduate nursing schools do not include content specific to the healthcare needs of adults with IDD as part of their generalist baccalaureate nursing education (Auberry, 2018). As people with IDD moved into the community over the past 60 years, the reduced experience and training of the health care providers, including nurses, around the healthcare needs of adults with IDD contributed to unexpected poorer health outcomes (Ervin et al., 2014). In addition, people with IDD in the community had less access to formal healthcare, and early symptoms of problems were often not recognized and managed in a timely manner (Janicki et al., 1999).

People with IDD have multiple distinctive patterns of medical comorbidities (often including cardiac malformations) associated with the underlying cause of the IDD (Coppus, 2013; Lauer & McCallion, 2015), increased medical complexity (Agency for Healthcare Research and Quality, 2019; Ervin et al., 2014; Lauer & McCallion, 2015), frequent reduction in capacity for communication (Auberry, 2018; Ervin et al., 2014; National Council on Disability, 2009), an entanglement of behavioral symptoms, and nontraditional presentation of symptoms for common conditions (Auberry, 2018; Ervin et al., 2014; Kyrkou, 2014; National Council on Disability, 2009). The result can be an arduous and intimidating patient case for even the most experienced of nurses. Without knowledge of the non-traditional signs and symptoms for common conditions frequently present in adults with IDD, conditions may worsen, possibly leading to failure to rescue (Agency for Healthcare Research and Quality, 2019).

Identification of major causes of mortality are important as this knowledge will create the basis for programs designed to reduce mortality (Heslop & Hoghton, 2019; Lauer et al., 2015). To date, very little has been done to identify the primary causes of death for those with IDD (Heslop et al., 2015; O'Leary et al., 2018). There is a critical need for information on causes of death and contributing factors in adults with IDD (Hemm et al., 2015). This is an important first step in efforts to decrease health disparities among this population.

I will use the National Minority Health and Health Disparities Research Framework (Alvidrez et al., 2019) to guide a cross-sectional study to examine common diagnoses and factors associated with mortality.
Administrative hospitalization data will be examined from the Healthcare Cost and Utilization Project National Inpatient Sample (Agency for Healthcare Research and Quality, 2021) for adults aged 18 and above, with and without IDD, hospitalized for any reason in the US in 2019. This study will identify base DRGs, ICD-10CMs and select factors associated with having IDD among hospitalized adults who expire.

Specific Aims

- **Aim 1**: Describe commonly reported base Diagnostic Related Groups (base DRGs) among adults with and without IDD who died during hospitalization 2019, controlling for biological, physical/built environment, and the healthcare system domains of influence on health outcomes at the individual level.
- **Subaim**: Among people who died during hospitalization, identify base DRGs that are more common in people with IDD than in people without IDD.
  - a. I will examine the base DRGs of adults with and without IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set, and compare the base DRGs controlling for age, gender, race, urbanicity of patient’s residence, and mean income of patient’s residence.
  - b. I will stratify the comparisons of base DRGs among adults with and without IDD by etiology and/or severity of IDD.
  - c. I will stratify the comparisons of base DRGs among adults with and without IDD by geographic area.
  - d. I will stratify the comparisons of base DRGs among adults with and without IDD by age.
- **Hypothesis**: The base DRGs for adults with IDD will be different from the base DRGs for adults without IDD.

- **Aim 2**: Describe commonly reported International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM) diagnoses preceding death during hospitalization for adults with and without IDD, stratified by etiology of IDD and/or severity of IDD and further stratified by geographical area of the US to assess for differences in patterns.
Subaim: Identify ICD-10-CM diagnoses that are more commonly associated with the most common base DRGs associated with mortality among people with IDD.

○ a. I will examine the recorded ICD-10-CM codes and base DRGs of adults with IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set. These ICD-10-CM codes and base DRGs will be stratified based on etiology and/or severity of IDD.

○ b. I will examine the recorded ICD-10-CM codes of adults with IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set for the US as a full set, and then also stratified by geographical area of the US.

○ c. I will examine the recorded ICD-10-CM codes of adults with and without IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set for the US as a full set, and then also stratified by age.

○ d. I will examine the recorded ICD-10-CM codes and base DRGs of adults with and without IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set, and then compare the prevalence of ICD-10-CM diagnoses between the groups.

Hypothesis: The recorded ICD-10-CM diagnoses and base DRGs for adults with IDD who expire in the hospital will be different from the recorded ICD-10-CM diagnoses and base DRGs for adults without IDD who expire in the hospital.

Aim 3: Describe hospital length of stay (LOS) and age at death, as well as select biological, physical/built environment, and the healthcare system domains that are associated with having IDD as compared to not having IDD with the same base DRG.

○ a. I will examine the above variables and base DRGs of adults with IDD who died in a community or tertiary hospital recorded in the HCUP/NIH 2019 data set.

○ b. I will stratify the comparisons of data describing adults with IDD and without IDD by etiology and/or severity of IDD, if Aim 1 analyses suggest heterogeneity within the adults with IDD.

○ c. I will stratify the comparisons of data describing adults with IDD and without IDD by geographic area, if Aim 1 analyses suggest heterogeneity within the areas.
○ d. I will stratify the comparisons of data describing adults with and without IDD by age, if Aim 1 analyses suggest heterogeneity within the areas.

○ Hypothesis: The patterns of hospital length of stay (LOS) and age at death, as well as select biological, physical/built environment, and the healthcare system domains of influence on health outcomes, including age, gender, race, urbanicity of the patient's residence, and the mean income of the patient's zip code will be different for adults with IDD as compared to adults without IDD with the same base DRG.

Results from this study will provide the data necessary to develop and test interventions targeting focused assessments of the signs and symptoms of diagnoses that commonly precede mortality among hospitalized adults with IDD. These assessments may subsequently prompt intervention that may decrease the likelihood of mortality as an outcome, one step in decreasing the disparities in health outcomes among this population.

**Review of Literature**

The purpose of this study is in the context of my overall goal to reduce avoidable deaths in adults with IDD. This section will examine concepts within this context, as well as concepts that allow the reader to form a frame of reference to other works in this field.

**Avoidable Deaths**

The concepts of amenable and preventable deaths rest on the idea that certain deaths could have been avoided if there had been more effective health care available (Office for National Statistics, 2019). Preventable deaths are any deaths which could have been avoided if higher quality public health interventions that focused on determinants of health had been in place, such as interventions to improve lifestyle choices, socioeconomic status and environmental factors, while amenable deaths are more focused on, and include deaths that could have been prevented if specific quality health care had been in place (Office for National Statistics, 2019). Amenable deaths are influenced by the level of technology and medical knowledge available preceding the time of the person's death, and access to care (Office for National Statistics, 2019). A death is considered avoidable if it is either preventable or amenable, or both (Office for National Statistics, 2019).
Avoidable death is much more common in adults with IDD, as high as 2.3–3.3 times more prevalent among people with IDD compared to the people without IDD (Glover et al., 2017; Heslop et al., 2014; Hosking et al., 2016; Shooshtari et al., 2020; Trollor et al., 2017). Importantly, existing definitions of amenable mortality do not include some significant and potentially treatable causes of mortality among people with IDD, such as urinary tract infections and aspiration, and thus likely do not encompass the true burden of avoidable mortality for people with IDD (Hosking et al., 2016).

Not all causes of death for people with IDD are avoidable. A list of avoidable conditions that cause mortality in this group that is familiar to nurses who specialize in care for people with IDD is called *The Fatal Four*, more recently referred to as *The Fatal Five*.

**The Fatal Four/Five**

Historically, IDD nurses have been taught about “the fatal four,” (1) constipation, (2) dehydration, (3) aspiration, and (4) seizures (Bailey, 2017), as causes of avoidable death in adults with IDD, but this teaching is not research based. Karen Green-McGowan, an eminent leader in the IDD field, is a developmental disabilities nurse who has worked with people with IDD since 1965 (IntellectAbility, 2021). McGowan has created a nationally recognized and widely-used health screening tool, the Health Risk Screening Tool, has authored many developmental disabilities training manuals for clinicians, founded the company IntellectAbility, served as president of the Developmental Disabilities Nurses Association (DDNA) for the 2015 and 2016 terms, (IntellectAbility, 2021), and has trained thousands of nurses on what she is now calling the fatal five (although it contains six elements; personal communication, March 23, 2021). Personal experiences of Green-McGowan, and the medical director she works with, led to a transition of the Fatal Four to now the Fatal Five, which contains six conditions: (1) sepsis, (2) aspiration, (3) bowel obstruction, (4) seizures, (5) dehydration, and (6) gastroesophageal reflux disease (GERD; personal communication, March 23, 2021). However, other educators continue to teach the more traditional fatal four (Relias, 2021).

While the Fatal Four/Five provides a beginning framework for nursing evaluation and intervention, inconsistency in teaching about the fatal four and the fatal five and lack of an evidence base for this teaching contributes to the challenges of nurses who work with and care for adults with IDD. Evidence-based practice is demanding data to support the suppositions of the experts’ personal experiences.
Mortality Studies Overview

Mortality in people with IDD has been studied from multiple viewpoints over the past 30 years. Some studies have considered mortality for certain etiologies of IDD such as autism (Bilder et al., 2013; Hirvikoski et al., 2016), Dravet syndrome (Cooper, M. S. et al., 2016), or Down syndrome (Eyman et al., 1991). Some studies break certain etiologies of IDD into further sub-populations and study, for example, mortality of people with Down syndrome who have dementia vs those without dementia (Coppus, A. M. W. et al., 2008), or mortality of people with IDD and epilepsy (Kiani et al., 2014; Morgan et al., 2001). Many studies separated the data for people with Down syndrome from the data for people with other types of IDD (Carter & Jancar, 1983; Chaney & Eyman, 2000; Oppewal et al., 2018; Puri et al., 1995). However, no available published studies to date stratify the causes of mortality among a cohort of people with many different etiologies of IDD. This is important as different syndromes that cause IDD also cause differing comorbid conditions, leading to divergent risks. For example, people with Down syndrome are at a higher risk for congenital cardiac defects, people with Fragile X syndrome are at higher risk for seizures, and people with Prader-Willi syndrome are at higher risk of hyperphagia and obesity (Prasher & Janicki, 2019).

Mortality and life expectancy are complementary, but different, concepts in population health that are often studied together (Woolf & Schoomaker, 2019). Life expectancy, one of the most used indicators for the overall health of a population (Luy et al., 2020), is an estimate of the number of years a person can expect to live (Bezy, 2012). Mortality, an indicator of the “unhealthiness” or “disease” of a population, is often considered a major inverse health indicator (Granados, 2012). The use of the indicator life expectancy is often based on the assumption that it directly reflects the associated mortality conditions of a certain population in a certain time period (Luy et al., 2020). Therefore, this review will include research based on both inextricably linked concepts.

Life Expectancy

Overall, the lifespan of people with IDD is increasing (Carter & Jancar, 1983; Chaney & Eyman, 2000). In the 1930s, the average lifespan for males with IDD was 14.9 years, and for females was 22 years (Carter & Jancar, 1983). By 1980, these lifespans had risen to 58.3 and 59.8 years respectively (Carter & Jancar, 1983).
The most recently published study shows an expected 61.1 year life expectancy for all types of IDD (Landes et al., 2021).

Studies have consistently shown that the lifespan for adults with IDD is lower than that of adults without IDD, however some studies show that this gap is reducing (Arvio et al., 2016). In 2016, Arvio and colleagues found that in Finland between the years of 1970 and 2012 the average lifespan of both people with IDD and people without IDD increased, and that the gap between the two decreased. In contrast, Emerson et al. (2014) showed that the lifespans for both groups have increased, without any change in the gap.

**Factors Affecting Life Expectancy**

Multiple factors have been identified as potential causes for reductions in the lifespan of adults with IDD. Recognizing these factors might enable nurses to identify signs and symptoms of a problem before they occur and may thus prevent the mortality associated with these problems/factors.

**Association between IDD Etiology and Severity and Life Expectancy.** Bittles et al. (2002) found a decrease in the lifespan of people who had IDD that was attributed to an underlying genetic disorder versus people with IDD due to a non-genetic reason ($p<0.0001$). This pattern has repeated in studies including people with Down Syndrome (Carter & Jancar, 1983; Hosking et al., 2016; Ng et al., 2017; Ohwada et al., 2013) but in contrast Chaney and Eyman (2000) reported that people with an onset of brain damage prior to birth had the lowest life expectancies, and non-chromosomal prenatal causes were associated with worse outcome than chromosomal prenatal causes. Cerebral palsy, a non-chromosomal etiology of IDD, was associated with the shortest lifespan (Landes et al., 2019; Reppermund et al., 2019).

Arvio and colleagues (2016) found a correlation between lifespan and severity of intellectual disability, with those with the most severe disability having the shortest lifespans. This correlation between severity of IDD and shorter lifespan has been seen in multiple studies (Bilder et al., 2013; Coppus et al., 2008; Hirvikoski et al., 2016; Landes et al., 2021; McCarron et al., 2015).

The increased mortality of people with IDD is a function of many factors over a person’s lifetime (Florio & Trollor, 2015). Understanding variations between etiologies of IDD and mortality may guide public health and preventive care efforts to reduce premature mortality (Landes et al., 2019).
Association between Gender and Life Expectancy Among Adults with IDD. Many studies report a decrease in lifespan for men with IDD as compared to women with IDD (Arvio et al., 2016; Florio & Trollor, 2015; Glover et al., 2017; McCarron et al., 2015; Ng et al., 2017). In contrast, Hirvikoski et al. (2016) and Ohwada et al. (2013) report that women with IDD had a shorter lifespan than men with IDD. Further, others report that gender does not affect lifespan in people with IDD (Landes et al., 2021; Shooshtari et al., 2020; Trollor et al., 2017). No studies were found that studied the effects of being transgender or specific etiologies of IDD that affect gender expression, such as Turner Syndrome. Additional study is necessary to discern any true association between gender and mortality.

Association between Comorbid Diagnoses and Life Expectancy Among Adults with IDD. Epilepsy (Coppus, 2013; Coppus et al., 2008; Hosking et al., 2016; Kiani et al., 2014; Ohwada et al., 2013) and dementia (Coppus, 2013) are predictors of lower lifespan in people with IDD. Although both dementia (Wolters et al., 2019) and epilepsy (Granbichler et al., 2017) reduce the life expectancy of a person without IDD, Hosking (2016) showed that the combination of IDD and epilepsy further lowered the already reduced anticipated lifespan.

Causes of Mortality Among People with IDD

Some recent studies report respiratory infections/respiratory diseases as the primary cause of mortality among people with IDD (Bilder et al., 2013; Coppus, 2013; Landes et al., 2021; Oppewal et al., 2018; Trollor et al., 2017), however other studies report circulatory system diseases as the most prevalent causes of death, with respiratory system diseases second (Glover et al., 2017; Hosking et al., 2016; Ng et al., 2017; Shooshtari et al., 2020). Respiratory causes of death are much more common in people with IDD than in people without IDD (Landes et al., 2021; Oppewal et al., 2018; Shooshtari et al., 2020), with death rates for pneumonia and aspiration pneumonia more than 10 times higher among adults with IDD than among adults without IDD (Hosking et al., 2016). Causes of mortality that are associated with high-risk lifestyle choices, such as using nicotine, alcohol or illicit drugs (Hosking et al., 2016; Janicki et al., 1999), and violence (Patja et al., 2001), are not contributing to mortality for adults with IDD.
Challenges of Studying Mortality and Life Expectancy Data with People with IDD

Despite the 30 years since mortality studies were recognized as important for people with IDD, efforts towards quality mortality studies in the US hampered by a lack of centralized database to collect health information of people with IDD in the US (Heslop et al., 2015a; Janicki et al., 1999; Landes et al., 2021). A centralized health information database is a common source of information in other countries such as England (Glover et al., 2017; Hosking et al., 2016; Kiani et al., 2014), Ireland (McCarron et al., 2015), Australia (Florio & Trollor, 2015), and Sweden (Hirvikoski et al., 2016). Incorrect coding on death certificates (Heslop et al., 2015; Heslop & Hoghton, 2019), including the issues of underreporting of IDD on death certificates (Dunwoodie Stirton & Heslop, 2018; Hosking et al., 2016; Tyrer & McGrother, 2009) and listing intellectual disability or associated condition as a cause of death (Dunwoodie Stirton & Heslop, 2018; Hoslop et al., 2015; Hosking et al., 2016; Landes et al., 2019b), also challenge researchers, leading some to call for abandoning the use of death certificate data when studying mortality in people with IDD (Hosking et al., 2016). This creates a challenge as many mortality studies for other populations are based on death certificate data, leaving researchers who are studying mortality among people with IDD searching for alternate resources to get similar data (Glover et al., 2017).

Additional challenges in mortality studies for people with IDD include the relatively small size of many studies, limiting the studies’ representativeness (Hosking et al., 2016; Lauer et al., 2015), the presence of long running longitudinal studies that limit the ability of the results to be contemporary (Hosking et al., 2016; McCarron et al., 2015), and lack of control populations from which to draw comparison conclusions (McCarron et al., 2015).

Commonly, mortality studies have presented causes of mortality in large categories of etiology, mostly based on ICD-10-CM coding chapters. While this is helpful, it does not help identify the granular cause of mortality. Limiting categories of cause of death to ICD-10-CM chapters severely restricts information about actual causes of death, making risk assessment and mitigation difficult.

Summary

Few studies to date compare data about mortality in a single hospitalized cohort of adults with and without IDD, and few studies further stratify mortality data by etiology of IDD, geography and age. Use of a
large database like the HCUP/NIH data set provides the power necessary to identify differences in conditions that occurred prior to death and will allow for comparison of factors between people with IDD and people without IDD.

Theoretical/Conceptual Framework

The NIMHD Minority Health and Health Disparities Research Framework (see Figure 1) will guide this study. This framework describes five domains of influence (Biological, Behavioral, Physical/Built Environment, Sociocultural Environment, Healthcare System) and four levels of influence (Individual, Interpersonal, Community, Societal), creating twenty unique sets of determinants which influence the outcomes of individual health, family/organizational health, community health and population health (National Institute on Minority Health and Health Disparities, 2017). Health disparities of individuals are seen as having social origins, which cannot be adequately addressed if only the individual’s factors are considered (Agurs-Collins et al., 2019). The determinants of health can be seen as direct factors leading to health outcomes or can be viewed as defined target areas towards which to design interventions to improve health and reduce disparities (Alvidrez et al., 2019).

This framework supports several underlying philosophies that are present in this project. The framework asserts that conducting research exclusively in one cell will result in research that does not address the intersectionality of the multiple determinants of health outcomes (Alvidrez et al., 2019; Duran & Pérez-Stable, 2019). Additionally, the framework supports the consideration of both biological and social determinants of health as they influence the health outcomes of minorities (Alvidrez et al., 2019; Duran & Pérez-Stable, 2019). Lastly, adults with IDD experience health disparities as defined by this framework.

This study will focus on the biological domain of influence at the individual level, focusing on biological vulnerability of people with IDD; the physical/built environment domain at the community level of influence, examining the influence of the rurality of the person’s residence and the median household income of the person’s zip code on their health outcome; and the health care system domain at the community level, examining the influence of the hospital size on the person’s health outcome. Considering aspects from multiple domains and with multiple levels of influence aligns with this framework (Alvidrez et al., 2019). The relationship between health determinants and health disparity outcomes outlined by Duran and Pérez-Stable
DIAGNOSES ASSOCIATED WITH INTELLECTUAL AND DEVELOPMENTAL DISABILITIES (2019; see Figure 2) will be used to guide the selection of dependent variables within the limitations of data that are available in the HCUP database.

Although only being formalized in 2019, the NIMHD Minority Health and Health Disparities Research Framework has already been the basis of multiple studies. It served as the framework for the Pain Coping Skills Training for African Americans with Osteoarthritis (STAART) trial, which aimed to improve pain coping through a phone-based intervention with African Americans who have been diagnosed with osteoarthritis (Allen et al., 2018).

Molleston and Bennett (2021) used the NIMHD Minority Health and Health Disparities Research Framework in the discussion of their secondary analysis of the Pediatric Health Information Data in their quest to describe the mortality, risk factors, and comorbidities of esophageal variceal bleeding in children. They identified potential causes of increased mortality in Black children with variceal bleeding as including environmental factors of reduced access to medical care or bias on the part of healthcare providers (Molleston & Bennett, 2021).

Bartels et al. (2020) used the NIMHD Minority Health and Health Disparities Research Framework in their study of lupus as they created metrics for retention in care and examined how race and other social determinants of health predicted lupus retention in care. They chose specific predictors of interest that were consistent with the framework including the individual-level factors of age, gender, race, ethnicity, and smoking status, and ZIP code, as well as the community-level factors of rural urban classification and neighborhood disadvantage quartile (Bartels et al., 2020).

The NIMHD Minority Health and Health Disparities Research Framework includes many points that will be applicable to this study, and, although new, has shown that it supports the research of a variety of studies supporting minority health.

**Methods**

**Design**

This study is a quantitative secondary analysis of administrative hospitalization data from the Healthcare Cost and Utilization Project (HCUP) National Inpatient Sample (HCUP-NIS; Agency for Healthcare Research and Quality, 2021) for adults aged 18 and above, with and without IDD, hospitalized for any reason

Sample

I will be using the 2019 HCUP dataset, which includes an approximately 20 percent stratified sample of discharges from all U.S. community (non-federal) acute care hospitals except from the states Alabama and Idaho (Agency for Healthcare Research and Quality, 2021). Just over 1/3 of Americans spend their final days in a hospital (Morbidity and Mortality Weekly Report, 2020) receiving nursing care, thus supporting the decision to use hospital data for this study. I will only be including data on patients aged 18 years and older when admitted. Limiting research to adults will reduce the influence of the early-life deaths expected due to the syndromal causes of IDD (Trollor et al., 2017).

Procedures and Data Acquisition

HCUP-NIS is maintained by the Agency for Healthcare Research and Quality in the United States. In order to purchase HCUP data I will create an account with the online HCUP Central Distributor and submit an application to purchase HCUP data. The members of my Dissertation Committee who will access HCUP data and I will complete the online HCUP Data Use Agreement (DUA) Training, which is about 15 minutes long, and sign the DUA. After registering for an account with the online HCUP Central Distributor I will complete the user profile and submit required DUA certification codes, completion dates, and the signed DUA. Once approved, I will download a zip file with the data from the online HCUP Central Distributor directly to a protected research drive which I have obtained from the school for this purpose (see below; Agency for Healthcare Research and Quality, 2021). Once my application is complete, the download should be available.

Measures

Data elements of interest that are contained in the HCUP-NIS include the first forty (40) ICD-10-CM diagnosis, the DRG, patient demographic characteristics (e.g., sex, age, race, median household income for ZIP Code), discharge status, length of stay, and severity and comorbidity measures (Agency for Healthcare Research and Quality, 2021).
Independent Variables

**Diagnosis of IDD.** The primary variable in this study is the presence or absence of a diagnosis of IDD. A participant will be considered to have IDD if their ICD-10-CM diagnosis codes list contains mild/moderate IDD (F70–71), severe/profound IDD (F72–73), unspecified IDD (F78–79), DiGeorge syndrome (D82.1), congenital iodine-deficiency syndrome (E00), classical phenylketonuria (E70.0), defects in glycoprotein degradation (E77.1), other disorders of glycoprotein metabolism (E77.8), Lesch-Nyhan syndrome (E79.1), other disorders of purine and pyrimidine metabolism (E79.8), disorders of psychological development (F84, F88–89), holoprosencephaly (Q04.2), other reduction deformities of brain (Q04.3), other specified congenital malformations of brain (Q04.8), tuberous sclerosis (Q85.1), Prader-Willi syndrome (Q87.11), or other chromosome abnormalities, not elsewhere classified (Q90-99) anywhere in the list. This is consistent with previous similar studies (Glover & Ayub, 2010; Ng et al., 2017).

**Dependent Variables**

**Base DRGs.** Base DRGs will be used to determine the person’s primary reason for being hospitalized. I will use the datapoint DRG in HCUP for this information and will transform the variable DRG into the base DRGs for some analyses.

**ICD-10-CM diagnoses.** While the base DRG will represent the reason for hospitalization, the ICD-10-CM diagnoses encompass the entire disease burden carried by the person. The data point ICD-10 Diagnoses will be used for this variable, which will label up to 40 of the person’s diagnoses.

**Hospital Length of Stay.** Length of stay, or the number of midnights spent admitted in the hospital, is reported in numeric form under data element LOS.

**Age at Admission/Death.** The patient’s age, which will be taken as age at admission, is in the data element AGE and is reported as age in years coded 0-124 years. Age at admission will be used as the person’s age at death.

**Gender.** The patient's gender is listed under the data element FEMALE, and is reported as (0) male, or (1) female. There is not a non-binary option.
**Race.** The patient’s race is reported under data element RACE, and includes coding (1) White, (2) Black, (3) Hispanic, (4) Asian or Pacific Islander, (5) Native American, (6) other. Importantly, for 2019, RACE contains missing values on over 3 percent of the records. I will check to see if the percentage of missing data differs for adults with and without IDD, and whether missingness of race is related to other patient characteristics. Decisions about whether to use race as a variable will be made with the committee after missingness has been evaluated.

**Urbanicity of patient's residence.** Location of patient’s residence is found at data element PL_NCHS. This is a six-category urban-rural classification scheme for U.S. counties is coded into the designations of (1) "central" counties of metro areas of >=1 million population, (2) "fringe" counties of metro areas of >=1 million population, (3) counties in metro areas of 250,000-999,999 population, (4) counties in metro areas of 50,000-249,999 population, (5) micropolitan counties, and (6) not metropolitan or micropolitan counties.

**Median income of the patient's zip code.** Median household income for a patient's ZIP code can be found at data element ZIPINC_QRTL. For 2019, the median incomes are defined as (1) $1 - $45,999; (2) $46,000 - $58,999; (3) $59,000 - $78,999; and (4) $79,000 or more.

**Geography.** Geographic regions of the hospital are available at the HCUP-NIS element HOSP_REGION, and are designated as Northeast, Midwest, South, and West, consistent with the designations defined by the US Census Bureau (United States, 1994).

**Unavailable.** Ideally, I would also like to evaluate a person’s living situation further, to evaluate if they are coming from a group home, a family home, section-8 housing, an assisted living facility, or a skilled nursing facility, however this information is not available in the dataset.

**Data Analysis**

All statistical analyses will be performed using SPSS ver. 28.0 statistical software (IBM, 2021) utilizing nationally weighted data. Data will be screened for accuracy including missing values, implausible values and outliers. To improve validity, data will be compared to previously established demographics, such as the expectation that it contains 1-2% of entries describing people with IDD (Maulik et al., 2011) and the average life expectancy in the US of 78.7 years old (Centers for Disease Control and Prevention, 2021). Histograms and box plots will be created for continuous variables and cell counts created for categorical variables. Descriptive
statistics and frequencies will be used to describe the sample. Committee member, Dr Crawford, will confirm all outputs.

**Stratification**

Groups will be stratified based on the appropriate specific aim. The primary stratification will use the ICD-10-CM to determine the presence of IDD. Many computations will be based on these two groups, those with and without IDD. As secondary stratification, the group of people with IDD will be further stratified based on etiology of IDD (Down syndrome, Rett syndrome, Fragile X syndrome, etc.) or intensity of IDD (mild, moderate, severe, profound) as the data is available in ICD-10-CM codes. Additional stratification factors, age at admission and geography, will be used to assess for confounders.

**Statistical Measures**

Many techniques will be used to obtain the information needed to fulfill the specific aims.

**Standardized Mortality Ratios (SMRs).** An SMR is the ratio of observed deaths in the population of interest over the expected deaths using the general population death rate (Kiani et al., 2014). It is a common measure used in mortality studies (Kiani et al., 2014) and it provides a useful way of assessing the extent of excess mortality in people with IDD compared with the general population (Heslop et al., 2015; Tyrer & McGrother, 2009). An SMR of 1.0 represents no difference in mortality between the population of interest and the general population, whereas a SMR of 2.0 represents a twofold increase of mortality in the population of interest (Tyrer & McGrother, 2009). An SMR of all base DRGs will be calculated with the number of observed deaths in people with IDD as the numerator and number of observed deaths in people without IDD as the denominator.

There is a warning against using an SMR with a small sample, which could introduce bias based on random variation (Heslop et al., 2015). However, if used with a sufficiently large database, SMR, in addition to average age at death, is the preferred statistic for studies with adults with IDD to ensure that more robust statistics emerge that will increase the comparability and validity of the study (McCarron et al., 2015).

Importantly, as part of the Data Use Agreement for HCUP-NIS (Healthcare Cost and Utilization Project, 2021), I acknowledged that the release or disclosure of information where the number of observations in any cell of tabulated data is ≤10 can increase the risk for personal identification of subjects. Therefore, I will not use
values of 1-10 in any text or tables. Decisions on how to manage data if any cells reveal these values will be made with my committee members.

**Assessment of Specific Aims.** The first specific aim identifies the most common DRGs that are associated with having IDD, stratified among the groups described above. This specific aim will be addressed using chi-square statistics. The chi-square table will be set as, along the X-axis, people with IDD and people without IDD. A secondary calculation will be made separating out the types and severity of IDD diagnosed, and then will be further stratified by geographic area. Along the Y-axis I will include the top ten most prevalent base DRGs for people with IDD, the top ten most prevalent base DRGs for people without IDD, and the base DRGs for each of the Fatal Five (Green-McGowan, personal communication, March 23, 2021; see Table 1). If chi-square statistics are significant, I will calculate SMRs, which will be used to compare results of this study with results of other studies.

For the Fatal Five topic of sepsis, I will include DRGs 864 through 872, which cover fevers, viral illnesses, bacterial and parasitic illnesses, and septicemia (Centers for Medicare & Medicaid Services, 2019). For the topic of aspiration, I will include DRGs 205 and 206, which include the ICD-10-CM diagnoses of (T17910A) gastric contents in the respiratory tract. DRGs 177, 178 and 179, respiratory infections and inflammations, will be included as they contain ICD-10-CMs for (J690) pneumonitis due to inhalation of food and vomit, and (J698) pneumonitis due to inhalation of other solids and liquids.

The third of the Fatal Five, bowel obstruction, will include the DRGs 388, 389 and 390, which includes diagnoses such as (K560) paralytic ileus, (K5641) fecal impaction, and (K5649) other impaction of intestine. DRGs 329, 330 and 331 will be considered as they cover major small and large bowel procedures, which may be an outcome of bowel obstruction.

Seizures, the fourth of the Fatal Five, are classified as part of DRGs 100 and 101. The group of ICD-10-CM codes that begin with the characters G40, which represents any recurrent seizure, will be included in this variable.

The fifth of the Fatal Five, dehydration, is classified as DRG 640 and 641, and contain diagnoses such as (E860) dehydration, (E861) hypovolemia, (E869) volume depletion, unspecified, and (E870) hyperosmolality and hypernatremia. However, this DRG also contains diagnoses such as (E162) hypoglycemia, unspecified,
(E46) unspecified protein-calorie malnutrition, and (R627) adult failure to thrive, which are not associated with dehydration. Therefore, a new variable will be recoded which will include only DRG 640 and 641 as main problems, along with ICD-10-CM E86 (dehydration) or E87.0 (hyperosmolality) as primary and secondary diagnoses. The final, and sixth of the Fatal Five, GERD, will include codes beginning with K21 (gastro-esophageal reflux disease) in ICD-10-CM codes.

It is anticipated that the conditions designated for the Y-axis (the top 10 from those with IDD, plus the top 10 from those without IDD, plus the Fatal Four) will duplicate, however the maximum number of conditions on the Y-axis will be 26. Duplications will be removed.

The second specific aim evaluates all the diagnoses associated with mortality for the stratified groups as discussed above. A chi-square will be used to assess the statistical significance of differences of prevalence of ICD-10-CM diagnoses among the populations as stratified. This chi-square will have the X-axis with the headings of people with IDD and people without IDD from the top 10 most prevalent base DRGs for people with IDD and the base DRGs for each of the Fatal Five. The Y-axis will include ICD-10-CM diagnoses, grouped as deemed appropriate after discussion with the committee. The goal of the groupings is to maintain the meaningfulness of the granularity of the data, while not missing correlations between the diagnoses that may only have only slight differences. Again, SMRs will be calculated for significant findings. The purpose for this aim is to see if any specific diagnoses are associated with any of the top ten most common base DRGs or the Fatal Five.

The comparison of patient and hospital factors that are associated with having IDD in the third specific aim will be assessed using ANOVA for continuous dependent variables and chi-square for dichotomous dependent variables.

**Precision Estimate**

The precision estimate is based on the second specific aim. The HCUP-NIS database contains information on more than 7 million inpatient records in 2019 (Agency for Healthcare Research and Quality, 2021). Estimating 77.9% as age 18 or over (Ogunwole et al., 2021), and 1% of the population as being identified as having IDD (Maulik et al., 2011), this leaves two groups; the group of adults without IDD, at over 5,453,000 participants; and the group of adults with IDD, at over 54,530 participants. Beta is set to 0.20, and
alpha to 0.05/26 = .0019, applying a Bonferroni correction to accommodate multiple hypothesis testing. As an illustrative calculation, prevalence is taken from Landes, Stevens and Turk (2021) who reported that the most common cause of mortality in both adults with and without IDD was heart disease. The prevalence of heart disease in people without IDD was 24.61%, and the prevalence for people with IDD was 15.61%. This is a difference of 0.09. Using a 2-group comparison with a Bonferroni correction, this study will be able to detect a difference of 0.7% (NCSS, 2021), suggesting that the dataset will provide more than sufficient precision to detect anticipated between-group differences.

Data Management

I will obtain a secure, encrypted, research drive from UMASS Medical School, and all research files associated with this study will be stored on this research drive. Committee members will be given access to the research drive as necessary. Any printed output will be kept in a locked cabinet and destroyed immediately at the conclusion of this study. All calculations will be performed using SPSS and will be confirmed by committee member, Dr Sybil Crawford. Consistent with the terms of the Data Use Agreement, to decrease risk of identification of persons or establishments I will not publish or report values of 1-10 in text or tables, nor will I publish or report data that can identify individual establishments directly or by inference. I will only allow access to HCUP Nationwide data to those who are authorized users and have signed a copy of the Data Use Agreement and completed the online Data Use Agreement Training Course.

Anticipated Challenges

Not Reaching Needed Sample Size

If the 2019 sample does provide the anticipated sample size, if the anticipated number of people in the sample are not adults, or the anticipated number of people do not have IDD, then the precision estimate will be recalculated. If the newly calculated precision estimate is over 1, then approaches such as using additional years data from 2018 and if necessary, 2017, will be discussed with the committee. I do not expect this to be a problem as the HCUP-NIS contains data from more than 7 million hospital stays each year across the US, and when weighted, it estimates more than 35 million hospitalizations.
Human Subjects Issues

The research protocol will be approved by the Institutional Review Board of the UMass Chan Medical School. I anticipate that the University of Massachusetts Institutional Review Board will determine that this proposed research is not human subject research.

Limitations

The secondary data analysis research method is very powerful but has many inherent limitations. Specifically, in research involving secondary data, the data was not collected to answer the specific research question posed by the researcher (Johnston, 2014), and thus not all aspects of my questions were able to be examined using this dataset. I had to use some data as a proxy for specific data. For example, I used the median income of the patient's zip code to reflect the individual's income. The HCUP data set includes 20% of hospital admissions, so data are not reflective of 100% of the hospitalized population. The most recent available data is from 2019, and thus won't reflect any changes that may have occurred in the past few years.

Due to the nature of hospital admissions and secondary data, we do not know how long the afflicting conditions were present prior to death. The conditions could have been chronic, short-term, or could have developed during the admission. This is important to nursing as knowing the timeframe of the conditions may impact the selection of potential interventions. This is essential knowledge when planning for potentially lifesaving nursing actions.

Use of SMRs in evaluation of mortality data, while the standard for many studies (Kiani et al., 2014), does have limitations. Notably, the choice of denominator, i.e. death rate of adults without IDD in the study sample vs death rate in the entire US, will affect the SMR. I have chosen to use the death rate of adults without IDD in the study sample as the denominator as (1) I will then have a comparison group for each of the study groups, and (2) the comparison groups will follow the same definitions for inclusion. I recognize that many other studies use other data sources to create the denominator for the SMRs, however, I am not able to identify an adequate comparison group.

Summary

This is a secondary data analysis of HCUP-NIS data to describe the most common factors associated with the diagnosis of IDD in adults who died during hospitalization. This study is designed with multiple
specific aims and sub-aims to answer many questions on the same large database, and to provide data that can be used as a comparison for other studies being conducted across the world. Using the granular perspective of DRGs and ICD-10-CMs, this study will specifically analyze data associated with The Fatal Four/Five, as well as data associated with the multiple domains of influence as defined by the NIMHD Minority Health and Health Disparities Research Framework to evaluate their influence on mortality in adults with IDD.

A comprehensive body of literature tells the story of people with IDD who are dying at an earlier age than people without IDD. This study, founded in the NIMHD Minority Health and Health Disparities Research Framework, will reveal the nature of this disparity in hopes that the next steps will start to reverse this outcome. Nursing science will directly benefit by identifying the most common factors associated with mortality in adults with IDD that are not as common in adults without, which will create target sites for interventions designed to reduce mortality and increase lifespan of this vulnerable group.
Figure 1.

The NIMHD Minority Health and Health Disparities Research Framework

<table>
<thead>
<tr>
<th>Domains of Influence (Over the Life course)</th>
<th>Levels of Influence*</th>
</tr>
</thead>
<tbody>
<tr>
<td>Biological</td>
<td>Individual</td>
</tr>
<tr>
<td></td>
<td>Interpersonal</td>
</tr>
<tr>
<td></td>
<td>Community</td>
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<tr>
<td></td>
<td>Societal</td>
</tr>
<tr>
<td>Biological</td>
<td>Biological Vulnerability and Mechanisms</td>
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<td></td>
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<tr>
<td>Behavioral</td>
<td>Health Behaviors</td>
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<td></td>
<td>Coping Strategies</td>
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<tr>
<td>Physical/Built Environment</td>
<td>Personal Environment</td>
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<tr>
<td>Sociocultural Environment</td>
<td>Sociodemographics</td>
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<td></td>
<td>Limited English</td>
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<td></td>
<td>Cultural Identity</td>
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<tr>
<td></td>
<td>Response to Discrimination</td>
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<tr>
<td>Health Care System</td>
<td>Insurance Coverage</td>
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<td></td>
<td>Health Literacy</td>
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<td>Treatment Preferences</td>
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<td></td>
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<tr>
<td>Health Outcomes</td>
<td>Individual Health</td>
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<td></td>
<td>Family/Organizational Health</td>
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<td>Community Health</td>
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<td>Population Health</td>
</tr>
</tbody>
</table>

Figure 2.

The Relationship Between Health Determinants and Health Disparity Outcomes

<table>
<thead>
<tr>
<th>Table 1.</th>
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</table>

ICD-10-CM diagnosis codes and DRGs to be considered equivalent to the Fatal Five diagnoses

<table>
<thead>
<tr>
<th>Seizures- defined by both DRGs and ICD-10-CMs</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>100 Seizures with mcc</td>
<td>G40.419 Other generalized epilepsy and epileptic syndromes, intractable, without status epilepticus</td>
</tr>
<tr>
<td>101 Seizures without mcc</td>
<td>G40.501 Epileptic seizures related to external causes, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.001 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, not intractable, with status epilepticus</td>
<td>G40.509 Epileptic seizures related to external causes, not intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.009 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, not intractable, without status epilepticus</td>
<td>G40.801 Other epilepsy, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.011 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, intractable, with status epilepticus</td>
<td>G40.802 Other epilepsy, not intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.019 Localization-related (focal) (partial) idiopathic epilepsy and epileptic syndromes with seizures of localized onset, intractable, without status epilepticus</td>
<td>G40.803 Other epilepsy, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.101 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, not intractable, with status epilepticus</td>
<td>G40.804 Other epilepsy, intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.109 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, not intractable, without status epilepticus</td>
<td>G40.811 Lennox-Gastaut syndrome, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.111 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, intractable, with status epilepticus</td>
<td>G40.812 Lennox-Gastaut syndrome, not intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.119 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with simple partial seizures, intractable, without status epilepticus</td>
<td>G40.813 Lennox-Gastaut syndrome, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.201 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, not intractable, with status epilepticus</td>
<td>G40.814 Lennox-Gastaut syndrome, intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.209 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, not intractable, without status epilepticus</td>
<td>G40.821 Epileptic spasms, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.211 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, intractable, with status epilepticus</td>
<td>G40.822 Epileptic spasms, not intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.219 Localization-related (focal) (partial) symptomatic epilepsy and epileptic syndromes with complex partial seizures, intractable, without status epilepticus</td>
<td>G40.823 Epileptic spasms, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.301 Generalized idiopathic epilepsy and epileptic syndromes, not intractable, with status epilepticus</td>
<td>G40.824 Epileptic spasms, intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.309 Generalized idiopathic epilepsy and epileptic syndromes, not intractable, without status epilepticus</td>
<td>G40.89 Other seizures</td>
</tr>
<tr>
<td>G40.311 Generalized idiopathic epilepsy and epileptic syndromes, intractable, with status epilepticus</td>
<td>G40.901 Epilepsy, unspecified, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.319 Generalized idiopathic epilepsy and epileptic syndromes, intractable, without status epilepticus</td>
<td>G40.909 Epilepsy, unspecified, not intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.401 Other generalized epilepsy and epileptic syndromes, not intractable, with status epilepticus</td>
<td>G40.911 Epilepsy, unspecified, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.409 Other generalized epilepsy and epileptic syndromes, not intractable, without status epilepticus</td>
<td>G40.919 Epilepsy, unspecified, intractable, without status epilepticus</td>
</tr>
<tr>
<td>G40.411 Other generalized epilepsy and epileptic syndromes, intractable, with status epilepticus</td>
<td>G40.A01 Absence epileptic syndrome, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.A09 Absence epileptic syndrome, not intractable, without status epilepticus</td>
<td>G40.A11 Absence epileptic syndrome, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.A19 Absence epileptic syndrome, intractable, without status epilepticus</td>
<td>G40.B01 Juvenile myoclonic epilepsy, not intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.B09 Juvenile myoclonic epilepsy, not intractable, without status epilepticus</td>
<td>G40.B11 Juvenile myoclonic epilepsy, intractable, with status epilepticus</td>
</tr>
<tr>
<td>G40.B19 Juvenile myoclonic epilepsy, intractable, without status epilepticus</td>
<td>R56.00 Simple febrile convulsions</td>
</tr>
<tr>
<td>R56.01 Complex febrile convulsions</td>
<td>R56.1 Post traumatic seizures</td>
</tr>
<tr>
<td>R56.9 Unspecified convulsions</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>GERD- defined by ICD-10-CMs</th>
<th>Aspiration- Defined by DRGs</th>
</tr>
</thead>
<tbody>
<tr>
<td>K21 Gastro-esophageal reflux disease</td>
<td>177 Respiratory infections and inflammations with mcc</td>
</tr>
<tr>
<td>K21.0 Gastro-esophageal reflux disease with esophagitis</td>
<td>178 Respiratory infections and inflammations with cc</td>
</tr>
<tr>
<td>K21.0 .... without bleeding</td>
<td>179 Respiratory infections and inflammations without cc/mcc</td>
</tr>
</tbody>
</table>
### Diagnoses Associated with Intellectual and Developmental Disabilities

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>K21.01</td>
<td>…… with bleeding</td>
</tr>
<tr>
<td>K21.9</td>
<td>Gastro-esophageal reflux disease without esophagitis</td>
</tr>
<tr>
<td>205</td>
<td>Other respiratory system diagnoses with mcc</td>
</tr>
<tr>
<td>206</td>
<td>Other respiratory system diagnoses without mcc</td>
</tr>
<tr>
<td>K21.99</td>
<td>Gastro-esophageal reflux disease with esophagitis</td>
</tr>
</tbody>
</table>

#### Constipation - Defined by ICD-10-CMs

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>K59.0</td>
<td>Constipation</td>
</tr>
<tr>
<td>K59.00</td>
<td>…… unspecified</td>
</tr>
<tr>
<td>K59.01</td>
<td>Slow transit constipation</td>
</tr>
<tr>
<td>K59.02</td>
<td>Outlet dysfunction constipation</td>
</tr>
<tr>
<td>K59.03</td>
<td>Drug induced constipation</td>
</tr>
<tr>
<td>K59.04</td>
<td>Chronic idiopathic constipation</td>
</tr>
<tr>
<td>K59.09</td>
<td>Other constipation</td>
</tr>
<tr>
<td>K59.3</td>
<td>Megacolon, not elsewhere classified</td>
</tr>
<tr>
<td>K59.1</td>
<td>Irritable bowel syndrome with constipation</td>
</tr>
</tbody>
</table>

#### Sepsis - Defined by DRGs

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>864</td>
<td>Fever and inflammatory conditions</td>
</tr>
<tr>
<td>865</td>
<td>Viral illness with mcc</td>
</tr>
<tr>
<td>866</td>
<td>Viral illness without mcc</td>
</tr>
<tr>
<td>867</td>
<td>Other infectious and parasitic diseases diagnoses with mcc</td>
</tr>
<tr>
<td>868</td>
<td>Other infectious and parasitic diseases diagnoses with cc</td>
</tr>
<tr>
<td>869</td>
<td>Other infectious and parasitic diseases diagnoses without cc/mcc</td>
</tr>
<tr>
<td>870</td>
<td>Septicemia or severe sepsis with mv &gt;96 hours or peripheral extracorporeal</td>
</tr>
<tr>
<td></td>
<td>membrane oxygenation (ECMO)</td>
</tr>
<tr>
<td>871</td>
<td>Septicemia or severe sepsis without mv &gt;96 hours with mcc</td>
</tr>
<tr>
<td>872</td>
<td>Septicemia or severe sepsis without mv &gt;96 hours without mcc</td>
</tr>
</tbody>
</table>

#### Dehydration - Defined by both DRGs and ICD-10-CMs

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>640</td>
<td>Miscellaneous disorders of nutrition, metabolism, fluids and electrolytes</td>
</tr>
<tr>
<td></td>
<td>with mcc</td>
</tr>
<tr>
<td>641</td>
<td>Miscellaneous disorders of nutrition, metabolism, fluids and electrolytes</td>
</tr>
<tr>
<td></td>
<td>without mcc</td>
</tr>
<tr>
<td>E86</td>
<td>Volume depletion</td>
</tr>
<tr>
<td>E86.0</td>
<td>Dehydration</td>
</tr>
<tr>
<td>E86.1</td>
<td>Hypovolemia</td>
</tr>
<tr>
<td>E86.9</td>
<td>Volume depletion, unspecified</td>
</tr>
<tr>
<td>E87.0</td>
<td>Hyperosmolality and hyponatremia</td>
</tr>
</tbody>
</table>

#### Bowel Obstruction - Defined by DRGs

<table>
<thead>
<tr>
<th>Code</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>329</td>
<td>Major small and large bowel procedures with mcc</td>
</tr>
<tr>
<td>330</td>
<td>Major small and large bowel procedures with cc</td>
</tr>
<tr>
<td>331</td>
<td>Major small and large bowel procedures without cc/mcc</td>
</tr>
<tr>
<td>388</td>
<td>Gastrointestinal obstruction with mcc</td>
</tr>
<tr>
<td>389</td>
<td>Gastrointestinal obstruction with cc</td>
</tr>
<tr>
<td>390</td>
<td>Gastrointestinal obstruction without cc/mcc</td>
</tr>
</tbody>
</table>
References

https://psnet.ahrq.gov/primers/primer/38/failure-to-rescue


https://www.ancor.org/resources/publications/links/fatal-four-major-health-issues-impact-individuals-intellectual-and


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DIAGNOSES ASSOCIATED WITH INTELLECTUAL AND DEVELOPMENTAL DISABILITIES


Summary of Changes from Proposal

Changes from proposal date 1/19/2022.

Change 1- My specific sims were amended to be much more clear and to more accurately describe my research goals.

Change 2- I limited the scope of the study to cover medical conditions associated with IDD, and less focus on personal factors such as age, gender and location of residence. These factors were included as covariates instead of dependent variables, as originally proposed. I moved the Fatal Four/Five to a more prominent position in the Specific Aims, and these became dependent variables.

Change 3- I did not include Standardized Mortality Ratios (SMRs) due to the small cell counts. Instead, I used Prevalence Ratios (PRs).
Diagnoses Associated with Intellectual and Developmental Disabilities in Adult Decedents:
A Secondary Analysis of Healthcare Cost and Utilization Project
National Inpatient Sample (HCUP/NIS) Data
Heather L. Briere

Intellectual and Developmental Disabilities (IDD)

“Intellectual disability is a condition characterized by significant limitations in both intellectual functioning and adaptive behavior that originates before the age of 22.”
(American Association on Intellectual and Developmental Disabilities, 2023)
My Personal Interest in this Topic

Introduced to the Fatal Four in previous work

Colloquial term, no empirical foundations

Spoke to the person who coined the term

Needed to find a way to answer the question

Fatal Four
- Constipation
- Dehydration
- Aspiration
- Seizures

Fatal Five plus One
- Sepsis
- Aspiration
- Bowel obstruction
- Seizures
- Dehydration
- Gastroesophageal reflux disease (GERD)

Importance

Adults with intellectual and developmental disabilities (IDD)

- Have poorer health outcomes (Ervin et al., 2014; Lauer & McCallion, 2015)
- Bear a higher percentage of preventable diseases (Glover et al., 2017)
- Tend to have more complex, multi-faceted medical conditions (Auberry, 2018)
- Suffer lower life expectancies (Arvio et al., 2016; Glover et al., 2017; Heslop et al., 2014; Landes et al., 2019; Lauer & McCallion, 2015; Maulik et al., 2011; O’Leary et al., 2018)
Importance

However, to date, very little has been done to identify the primary causes of death for those with IDD (Heslop et al., 2015; O'Leary et al., 2018).

Nurses rely on accurate, specific, mortality information to design and administer programs and to care for patients to reduce the excess mortality of adults with IDD.

My Goals

Not just to provide support for or against The Fatal Four/Five(Six)

Which diagnoses are related to mortality at a higher rate for adults with IDD than for adults without IDD

By identifying the diagnoses related to mortality hope to identify factors nurses could try to mitigate to head off the negative mortality outcome
Purpose

The purpose of this study is to describe the diagnoses assigned to adults with IDD preceding their death that occurred at a higher rate than diagnoses assigned to adults without IDD preceding their death.

How Hospitals Document Diagnoses- A Brief Primer

1. The provider assigns a diagnosis at the bedside.
2. This diagnosis is “coded,” or translated into an International Classification of Diseases (ICD-10-CM) code that changes the diagnosis from a word to a string of letters and numbers.
3. The ICD-10 codes are “ranked” for each patient depending on their importance of that code for that person’s admission.
4. Diagnostic Related Groups (DRGs) use the first couple of ICD-10s for each patient. They are further coded into groups that establish the main reason that the person was admitted into the hospital.
Specific Aims

The aims of this study

1) to describe the commonly reported base diagnostic related groups (base-DRGs) preceding death among decedents with and without IDD who died during hospitalization 2019,

2) to determine which base-DRGs had a higher prevalence rate among adults with IDD than among adults without IDD, controlling for age, gender, race, urbanicity of person’s residence, US census division of hospital, and mean income of person’s zip code, and

3) to use the base-DRGs and ICD-10-CMs to examine the conditions of the Fatal Four/Five as conditions of concern preceding death in decedents with and without IDD.

Conditions of Concern

I defined a condition of concern as one that was

- represented in the most prevalent base-DRGs
- seen at a higher prevalence among decedents with IDD than decedents without IDD
- was clinically related to and supported by a prevalent ICD-10-CM
NIMHD Minority Health and Health Disparities Research Framework

The NIMHD Minority Health and Health Disparities Research Framework is a multi-dimensional model that supports the use of a wide array of health determinants relevant to understanding and addressing minority health and health disparities across the lifespan.

(National Institute on Minority Health and Health Disparities, 2017)

Methods: Secondary data analysis of the Healthcare Cost and Utilization Project National (Nationwide) Inpatient Sample (HCUP-NIS) data from 2019

HCUP-NIS

- The largest collection of longitudinal hospital care data in the United States.
- Includes an approximately 20 percent stratified sample of discharges from all U.S. community (non-federal) acute care hospitals except from the states Alabama and Idaho (Agency for Healthcare Research and Quality, 2021).
- When weighted, represents more than 35 million hospital admissions annually.
Inclusion Criteria

- Adults aged 18 and older at the time of admission to the hospital
- Died during hospitalization between January 1, 2019 and December 31, 2019

IRB exemption

This study was approved by the Institutional Review Board of UMass Chan Medical School, which deemed the study exempt from human subjects review due to its being a secondary analysis of existing data with de-identified data only.

Analyses

All calculations and analyses were performed using SPSS 28.0 statistical software and Excel utilizing nationally weighted data with weights provided by HCUP-NIS.

Independent variable

- Decedents with IDD
- Decedents without IDD

Dependent variables

- base-DRG
- ICD-10-CM

Covariables

- Age
- Gender
- Race
- Urbanicity of person’s residence
- Mean income of person’s zip code
Specific Aim 1: Describe the commonly reported base-DRGs preceding death among decedents with and without IDD

Ran 2X2 frequency distributions and chi-square statistics comparing the independent variables with dependent variables

Ran prevalence ratios (PR) on the base-DRGs that had cell counts above 10.

Result: Of the 46 base-DRGs evaluated (had a cell count over 10), 15 were statistically more prevalent in decedents with IDD.

Top Three Most Prevalent base-DRGs

<table>
<thead>
<tr>
<th></th>
<th>DECEDECTS WITH IDD</th>
<th>DECEDECTS WITHOUT IDD</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>SEPTICEMIA OR SEVERE SEPSIS WITHOUT MECHANICAL VENTILATION &gt;96 HOURS</td>
<td>SEPTICEMIA OR SEVERE SEPSIS WITHOUT MECHANICAL VENTILATION &gt;96 HOURS</td>
</tr>
<tr>
<td>2</td>
<td>RESPIRATORY SYSTEM DIAGNOSIS WITH VENTILATOR SUPPORT</td>
<td>RESPIRATORY SYSTEM DIAGNOSIS WITH VENTILATOR SUPPORT</td>
</tr>
<tr>
<td>3</td>
<td>SEPTICEMIA OR SEVERE SEPSIS WITH MECHANICAL VENTILATION &gt;96 HOURS</td>
<td>OTHER FACTORS INFLUENCING HEALTH STATUS*</td>
</tr>
</tbody>
</table>

Note. * could include stroke, coma, homicidal ideations, medication complications, psychiatric examination, examination after sexual assault, overdose, or many other possibilities
**Slide 17**

15 most prevalent base-DRGs statistically more prevalent in decedents with IDD.

1. SEPTICEMIA OR SEVERE SEPSIS WITHOUT MV >96 HOURS WITH MCC OR WITHOUT MCC
2. RESPIRATORY SYSTEM DIAGNOSIS WITH VENTILATOR SUPPORT
3. SEPTICEMIA OR SEVERE SEPSIS WITH MV >96 HOURS
4. INFECTIOUS AND PARASITIC DISEASES WITH O.R. PROCEDURE WITH MCC, WITH CC, WITHOUT CC/MCC
5. SIMPLE PNEUMONIA AND PLEURISY WITH MCC, WITH CC, OR WITHOUT CC/MCC
6. RESPIRATORY INFECTIONS AND INFLAMMATIONS WITH MCC, WITH CC, OR WITHOUT CC/MCC
7. SEIZURES WITH MCC, OR WITHOUT MCC
8. MISCELLANEOUS DISORDERS OF NUTRITION, METABOLISM, FLUIDS AND ELECTROLYTES WITH MCC, OR WITHOUT MCC
9. G.I. OBSTRUCTION WITH MCC, WITH CC, OR WITHOUT CC/MCC
10. ACUTE LEUKEMIA WITHOUT MAJOR O.R. PROCEDURE WITH MCC, WITH CC, OR WITHOUT CC/MCC
11. OTHER DISORDERS OF NERVOUS SYSTEM WITH MCC, WITH CC, OR WITHOUT CC/MCC
12. OTHER VASCULAR PROCEDURES WITH MCC, WITH CC, OR WITHOUT CC/MCC
13. STOMACH, ESOPHAGEAL AND DUODENAL PROCEDURES WITH MCC, WITH CC, OR WITHOUT CC/MCC
14. COMPLICATIONS OF TREATMENT WITH MCC, WITH CC, OR WITHOUT CC/MCC
15. KIDNEY AND URINARY TRACT NEOPLASMS WITH MCC, WITH CC, OR WITHOUT CC/MCC

**Slide 18**

Most prevalent ICD-10-CMs

<table>
<thead>
<tr>
<th>Decedents with IDD</th>
<th>Decedents without IDD</th>
</tr>
</thead>
<tbody>
<tr>
<td>Do not resuscitate status</td>
<td>3315 (64.24%)</td>
</tr>
<tr>
<td>Encounter for palliative care</td>
<td>2760 (53.49%)</td>
</tr>
<tr>
<td>Acute respiratory failure with hypoxia</td>
<td>2430 (47.09%)</td>
</tr>
<tr>
<td>Sepsis, unspecified organism</td>
<td>2400 (46.51%)</td>
</tr>
<tr>
<td>Acute kidney failure, unspecified</td>
<td>1910 (37.02%)</td>
</tr>
<tr>
<td></td>
<td>401465 (59.55%)</td>
</tr>
<tr>
<td>Encounter for palliative care</td>
<td>344765 (51.14%)</td>
</tr>
<tr>
<td>Acute respiratory failure with hypoxia</td>
<td>260825 (38.69%)</td>
</tr>
<tr>
<td>Acute kidney failure, unspecified</td>
<td>255010 (37.83%)</td>
</tr>
<tr>
<td>Acidosis</td>
<td>233195 (34.59%)</td>
</tr>
</tbody>
</table>
Specific Aim 2: Determine which base-DRGs had a higher prevalence rate among adults with IDD than among adults without IDD, controlling for age, gender, race, urbanicity of person’s residence, and mean income of person’s zip code.

Continuing with the 15 base-DRGs that were more common (had a PR >1.0) in decedents with IDD and had a statistically significant difference in prevalence, I ran a separate binomial logistic regression for each base-DRG and ICD-10-CM of interest, including adjustments for covariates to determine the association of having IDD on probability of having the base-DRG or ICD-10-CM at time of death.

The base-DRGs with the highest odds ratios for having an IDD diagnosis:

- Seizures (3.46, 95% CI [2.69, 4.45])
- GI obstruction (3.39, 95% CI [2.47, 4.65])
- Respiratory infections and inflammations (3.17, 95% CI [2.62, 3.83])
Specific Aim 3: Use the base-DRGs and ICD-10-CMs to examine the conditions of the Fatal Four/Five as conditions of concern preceding death in decedents with and without IDD.

Created variables that defined the Fatal Four/Five diagnoses

Performed parallel calculations to those described for specific aims one and two.

Looked for concordance between the base-DRGs, the Fatal Four/Fives, and the 15 most prevalent diagnoses on the ICD-10-CM list for decedents with IDD.

---

Three most prevalent ICD-10-CM diagnoses are the same: Do not resuscitate status; Encounter for palliative care; Acute respiratory failure with hypoxia.

After that-

**Decedents with IDD**
- Epilepsy
- Pneumonitis
- Sepsis
- Acidosis
- GERD
- Hypothyroidism
- Hypernatremia

**Decedents without IDD**
- Hyperlipidemia
- Atherosclerotic heart disease
- Essential hypertension
- Personal history of nicotine dependence

People with IDD tend to not smoke, and CAD is associated with smoking
Discussion

A new set of fatal conditions is proposed to assist nurses in reducing preventable deaths in decedents with IDD.

Two categories are proposed

**IDD Concerning Conditions** and **IDD Contributing Conditions**

### IDD Concerning Conditions
- Dehydration
- GI obstruction
- Respiratory infection
- Seizures
- Sepsis

### The IDD Contributing Conditions
- Aspiration
- Constipation
- GERD
Implications

Clinical- Nurses should be monitoring for the IDD Contributing Conditions and Concerning Conditions, and when identified to intervene to potentially prevent mortality in this vulnerable group.

Educational- Staff and supportive personnel in group homes with adults with IDD should also be made aware of the IDD Contributing and Concerning Conditions. Educators working with nursing students and nurses aides/medical assistants can also add this content to their curriculum.

Policy- Shift funding to research on this topic and investigate the creation of performance standards specifically for patient care of adults with IDD in health care facilities accredited by the Joint Commission, ensuring better quality of care for all.

Slide 26

<table>
<thead>
<tr>
<th>Strengths</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dataset provides a large number of cases from the large geography of the USA, increasing the representativeness of the study</td>
</tr>
<tr>
<td>Data from 2019 provides contemporary, pre-COVID-19 data</td>
</tr>
<tr>
<td>There is a built-in comparison set since the data includes all hospitalized decedents from 2019, not just decedents with IDD.</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Limitations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Researcher’s inability to confirm data or obtain data from variables that were not originally collected</td>
</tr>
<tr>
<td>HCUP-NIS restriction to not report any cells with values of 10 or fewer</td>
</tr>
<tr>
<td>Limited data- unable to consider other factors that have been shown to affect mortality.</td>
</tr>
<tr>
<td>Analyses did not adjust for multiple hypothesis testing, in order to maximize the ability to detect or discover possible associations to be studied further.</td>
</tr>
<tr>
<td>It may be challenging for hospitalists to identify and diagnose certain types and severities of IDD in the acute-care setting, thus biasing the data towards null.</td>
</tr>
</tbody>
</table>
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Heather L. Briere
Next Steps

Additional Research:

- Confirmatory research for the Concerning and Contributing Conditions using different datasets and different types of research.
- Why are these Concerning and Contributing Conditions more prevalent in decedents with IDD?
- Is early identification and intervention to alleviate these conditions among adults with IDD associated with decreased mortality during hospitalization?
Dissemination Plan

The primary description of this dissertation work was submitted as a manuscript on March 17, 2023 to the American Journal on Intellectual and Developmental Disabilities for review and consideration for publication.