Diffuse Intrinsic Pontine Glioma (DIPG); A Qualitative Study

Principal Investigator: Amy Deaner, (Nursing Student in BSN program)
Faculty Mentor; Kathy Zimmerman, PhD-c, MSN, APRN, FNP-BC, APHN-BC, CCH
Austin Peay State University, Clarksville, Tennessee

Abstract

An overwhelming experience for parents is the diagnosis of cancer for their child, especially a type that is rare, and offers few treatment options. The leading cause of cancer related deaths in children is brain cancer. While treatment and survival rates for pediatric cancer have improved over the past 30 to 40 years, there remain few effective treatments and no advancement in survival rates for children diagnosed with Diffuse Intrinsic Pontine Glioma (DIPG). DIPG comprises 10 to 15 percent of all pediatric brain tumors and an estimated 200 to 400 children, with a median age of six years, are stricken every year in the United States alone. Likewise the median survival rate is only nine months. There is a scarcity of research to indicate a genetic or environmental cause. Consequently recent research suggests that DIPG tumor formation may be linked to brain development. The purpose of this study was to better understand parents’ experience caring for a child with DIPG and identify any potential warning signs in early development. In depth interviews with participants provided information on their lived experience and what it is like to have a child diagnosed with DIPG. Three main themes emerged as a result of the study. Parents explained that quality of life for their child was most important, followed by ensuring their child died with dignity. Lastly, being mindful of respecting the child’s willingness to participate in treatment was a dominant theme. The hope is that this study will help to bring public awareness of the existence of this lethal cancer, in an effort to foster more research to develop a cure.

Introduction

What is DIPG?

Diffuse Intrinsic Pontine Glioma (DIPG) is a pediatric brain tumor located in the brainstem, specifically the pons. This area of the brain controls life functions such as breathing and heart rate, and houses the nerves involved in purposes of seeing, hearing, walking, talking, and eating. A DIPG tumor is not a solid mass, but a widely disseminated group of cells that makes surgical removal impossible. One way to visualize DIPG is to imagine pepper being sprinkled into an unset bowl of gelatin. As a result of its location and diffuse nature, DIPG is considered the most lethal of all brain tumors (Wagner et al., 2006). Not only is DIPG swift in its progression, it eventually robs its victim of the ability to move or communicate, while sparing the child’s cognitive abilities. In other words, the child is completely aware of what is happening while being trapped in a body they can’t manipulate. Ultimately, the tumor takes the child’s life when breathing and heart rate can no longer be maintained (National Cancer Institute, 2008).

DIPG Statistics:

As survival rates for pediatric cancer have increased over the last 30 to 40 years, there has been no improvement in mortality for children diagnosed with DIPG. Comprising 10 to 15 percent of all brain tumors in children, DIPG affects an estimated 200-400 children annually in the United States alone (Foer & Fisher, 2011). To put this in perspective, a child diagnosed with DIPG today faces the same prognosis as a child diagnosed 40 years ago. There is still no effective treatment and no chance of survival. Only 10% of children with DIPG survive for 2 years following their diagnosis, and less than 1% survives for 5 years. The median survival is 9 months from diagnosis (Johung & Monje, 2016). Please see visual provided by the Michael Mosier Defeat DIPG Foundation below. Permission granted.

Methodology

• Qualitative Interpretive Phenomenology
  • This method provides description and interpretation of participants lived experience.

Participants

A purposeful sample of parents were recruited from a flyer provided to the DIPG@yahoo-groups.com via email. Inclusion criteria consisted of parents who had lost a child as a result of DIPG within the last ten years, spoke English as a first language, and were at least 18 years of age.

Research Question

What is the lived experience of parents of a child diagnosed with Diffuse Intrinsic Pontine Glioma (DIPG)?

Data Collection

Semi-structured scheduled telephone interviews

Analysis

Interviews transcribed with careful attention to experiential descriptions and thematically analyzed through writing and re-writing

Additional Data Sources

Reflective Journal

Guiding Questions

• Please describe the moment your child was diagnosed with DIPG.
• How long did your child survive from diagnosis?
• Can you explain the cost of having a child with DIPG? (financial, emotional, marital, etc.)
• What types of support did your family have after the DIPG diagnosis?

Research Results

• Three main themes emerged from data analysis.
  1. Maintaining Quality of Life for the child
  2. Allowing child to die with dignity
  3. Respecting the child’s willingness to participate in treatment
• There is a lack of overall knowledge among Healthcare providers regarding DIPG treatment options at diagnosis. This includes ineffective communication among specialists in Pediatric Oncology performing clinical trials within the United States and abroad.
• There is a significant lack of resources provided to DIPG families upon diagnosis.
• This includes inaccurate data at times relative to prognosis, a lack of child-life specialists within institutions, and a reluctance to involve palliative care teams in a timely manner.
• There is a strong need for guidance and information to newly diagnosed DIPG families. An online resource location is highly recommended. This can be in the form of an online portal and a printed resource guide that can be provided to all DIPG families. This resource should include any and all clinical trials that their child may qualify for both at diagnosis and throughout treatment. This should also include information related to tumor donation and end of life decision making.

Implications

• Enhanced communication among Pediatric Oncologists involved in clinical trials.
• Formulation of a Resource Guide for DIPG families at time of diagnosis.
• Adequate staffing of child-life specialists in all diagnosing institutions.
• Education of DIPG to include treatment options and prognosis provided to all Healthcare providers.
• Earlier introduction of palliative care teams to include hospice care to all DIPG families.
• Discussion of possible tumor donation prior to end of life stages to all DIPG families.
• Logistical removal of donated tumor within 12 hours to ensure live tissue protocols are maintained, regardless of time or date of death, or geographic location.
• Increased awareness of DIPG within the Medical research community to procure the much needed financial resources in an effort to develop a cure.

Discussion

• While each DIPG is different in the genetic mutation it manifests, the presentation of its existence is vastly similar in children at time of diagnosis. Children who were healthy just days before, often present with vision problems, balance and gait issues, and severe behavioral changes. While parents try to make sense of the sudden onset of symptoms, the child continues to decline until he or she can no longer compensate. Many times an MRI is the diagnostic tool that uncovers the tumor hiding deep within the child’s brainstem. Parents are left reeling with the news that not only does their child have a brain tumor, but there is no surgical option for this one, called DIPG, and that care is considered palliative as there is no cure.
• Palliative options include radiation, which may slow the growth of the tumor, buying more time, that is labeled a “honeymoon”. Providers are careful to explain that not all children will experience this “honeymoon” and that many times, the radiation speeds up the tumor’s progression. They are also quick to point out that even if the tumor responds to radiation, it will eventually grow back, and they cannot re-irradiate. Occasionally, a clinical trial will be offered, depending on where the family lives in the United States, but these are few and require specific qualifying criteria to get into.
• Needless to say, a DIPG diagnosis is a death sentence that parents are powerless to stop. The lives they lived before diagnosis abruptly halt, and the future focuses on quality of life for their child, however long that life may be. Among the participants in this study, the median time frame from diagnosis of DIPG to the child’s passing, was ten months.
• Five families were selected for this study. They are the parents of three girls and two boys with a median age of seven years old at diagnosis of DIPG. Many themes were uncovered throughout the research. Maintaining quality of life, allowing their child to die with dignity, and respecting the child’s willingness to participate in treatment are the most relevant. Each parent expressed feelings of guilt and anger, frustration and powerlessness. Most of all, the parents wished they had known sooner that DIPG was the reason for the sudden change in their child. Lastly, all five families expressed how unfair this cancer can be and the enormous toll it has had on their daily lives.

References


Pediatric Cancer 5 - Year Survival Rates

Acknowledgements

• The principal investigator would like to thank the parents who volunteered to participate in this research. Your strength is to be admired and your candor is so appreciated. Together, we have brought awareness to DIPG in an effort to foster better resources for DIPG families, and more research to develop a cure.
• The principal investigator would like to thank former Research instructor and mentor, Mrs. Kathy Zimmerman. Your guidance and support were instrumental in the conception of this first research project.
• The principal investigator would like to sincerely thank the IRB at APSU, specifically Dr. Jorniann Butterfield, for allowing her to pursue this research.
• The principal investigator would like to thank Sigma Theta Tau International Honor Society of Nursing for giving her the opportunity to present this research.