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DIPG Symptom Management and End of Life Care (Physicians/Providers)



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DIPG Symptom Management (Physicians/Providers)

The symptoms listed below are common symptoms we see in our DIPG patient population that increase from the time of progression to the end of life affecting their quality of life. Listed below are explanations and suggestions for intervention. The primary care team should only use this as a general guide for potential options for therapy/intervention.

Constipation

Possible Etiologies: medications including opioids and steroids and/or neurologic secondary to tumor effect.

Recommended Interventions: osmotic agents (i.e. Lactulose ®, Miralax ®, Colace ®, Docusate ®, and magnesium citrate) and stimulants (i.e. Senna ®, Bisacodyl ®) or glycerin suppository. If thought to be related primarily to opioids, methylnaltrexone may be considered; however, administration is Sub Q and may induce distress to the patient.

Headache and other neurologic symptoms

Possible Etiologies: peritumoral edema, tumor enlargement resulting in hydrocephalus

Recommended Interventions:

Headache: steroids and/ or opioids

Seizures: rare in DIPG patients (< 20%). Anti-convulsants, specifically Keppra ® as this medication has limited interactions with chemotherapy; benzodiazepine at the bedside readily available is especially important in the end of life setting.

Dysconjugate gaze resulting in visual disturbances: alternating patching eyes, dark glasses, dim lights

Sensitivity to noise: ear plugs, calming music

Pain

Possible etiologies: tumor burden, arthritic pain due to steroids, abdominal pain secondary to constipation

Recommended Interventions: opioids (short acting and long acting; fentanyl patches if difficulty swallowing medications and no IV access), Tylenol ®, heat packs, massage therapy, NSAIDs with caution if on chemotherapy.

Nausea/Vomiting

Possible Etiologies: tumor burden, hydrocephalus, gastritis secondary to steroids, opioids

Recommended Interventions: Anti-emetics (i.e. Ondansetran, Promethazine, Benadryl ®, Ativan ®, Scopolamine Patch), peppermint and ginger may provide comfort. Surgical intervention with placement of shunt is case dependent.

Anxiety

Although the symptoms are primarily due to tumor progression, patients remain cognitively intact and depending the age of the patient, may be aware of the anticipated events. We recommend to encourage open, honest, and developmentally appropriate conversations with the patient. In some cases, withholding information may increase anxiety in the patient. We try

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to offer opportunities to share in decision making when appropriate. Medications and psychosocial support are beneficial.

Sleep disturbance/Fatigue

Possible etiologies: tumor burden, therapy related, activity related, progression of disease

Recommended Interventions: Melatonin, Ambien® (age dependent), Trazadone, or Remeron® for sleep disturbance. Stimulants for wakefulness (i.e. Methylphenidate).

Key is to balance comfort and sedation versus wakefulness.

Dysarthria/ communication difficulties (verbal and nonverbal)

Most patients remain cognitively intact while the disease progression evolves to a “locked-in-syndrome” resulting in motor impairment. Furthermore, the terminal phase may result in disturbance of consciousness.

Recommended Interventions: anticipatory guidance to the patient and parents early in the disease course and possible early referral to occupational/ speech therapy services for education regarding communication tools (i.e. I pads, picture/ word boards). If patients and parents are aware of the anticipated communication difficulties, they may choose to initiate legacy memory making early in the disease process.

Advanced Care Planning

It is recommended that you continually discuss the patient and families goals of care given the poor chance for survival with DIPG and the earlier this discussion is had, the better. These conversations will evolve over the course of the disease and are not static. Waiting may risk the patient not being able to effectively communicate their wishes. If the patient and family wish to be at home for end of life care, state-specific documents (DNR, MOST, POST, AND, etc.) will need to be in place.

Dysphagia

Possible etiologies: tumor location

Recommended Interventions: add thickener to thin liquids, small bites. Some providers may proceed with Nasogastric tube or G-tube placement, depending on patient/family goals of care. Some families/care providers opt to allow small amounts of oral feeding for pleasure even in the setting of possible aspiration.

Nutrition

Patients usually self-limit their oral intake with disease progression as part of the natural dying process and the body's need for food and hydration decreases.

Aspiration and choking are concerns with DIPG progression due to dysphagia; however, some patients and families choose to continue to have some oral intake

Artificial nutrition (TPN) and Intravenous fluids may actually cause more distress with air hunger due to accumulation of the fluid in the lungs.

Urinary Retention

Possible Etiologies: tumor burden, secondary to opioids

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Recommended Interventions: trial of Ditropan, in and out straight catheterizations versus foley placement.

Secretions

Possible etiologies: tumor progression resulting in sleep and difficulty in swallowing secretions

Recommended Interventions: anti-cholinergics (i.e. Scopolamine Patch, glycopyrrolate, diphenhydramine). Some patients and parents find suction machines are helpful.

Dyspnea/Air hunger

Possible Etiologies: pending brain herniation

Recommended Interventions: opioid medications and anxiolytic medications, blow by oxygen for comfort.

Immediate Dying Process

The dying process is a unique experience for each DIPG patient. This section will attempt to provide general guidance on what to expect as the person enters the final days of life. It is important to emphasize that the signs and symptoms discussed do not occur with each person and may not occur in the sequence presented. Each patient follows his/her own timeline and we will attempt to provide you with the information needed to help the patient and family during this difficult time. For some patients, the timeline may be in months while for other patients it may be in days. Our goal is to help you, the provider, make the patient and family as comfortable as possible no matter the timeline. The involvement of a palliative care team and/or hospice program, if available, is ideally recommended to provide needed support to everyone involved.

Disorientation

Patients commonly become confused about time, place and the identity of the people around them. This may worsen as the patient enters the final days and hours of life. It is important that families and medical staff speak softly and clearly to lessen any negative reaction from the patient. Medications can be used to assist the patient if the symptoms are consistent with delirium and it is believed that treatment may improve their quality of life.

Decrease in food and fluid intake

Both food and fluids are necessary for life but become unnecessary in the time prior to death. This is a very difficult topic for patients, families and physicians. As a patient begins the dying process, his/ body's need for sustenance declines and his/her appetite and thirst decrease. We encourage parents to not force the patient to eat or drink and offer food and drink in small amounts for comfort. Intravenous fluids and parental nutrition may exacerbate respiratory distress and we present to the families that the risks outweigh the benefits during this time.

Sleeping

As patients enter the last stages of dying, they will often begin sleeping more as their body begins to shut down. At times, sleepiness may be due to medications that are prescribed for symptoms such as pain or anxiety. We encourage families to continue to spend time with their loved ones during this time as the patient may be able to hear them speak and derive some comfort from this interaction.

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Congestion

The patient may have increased secretions and congestion during the last stages of the dying process. This may include gurgling and other sounds that may be difficult for the family to hear. Use of medications to decrease secretions as well as limiting fluid intake (both by mouth and intravenous) is often necessary. It is important to relay to the family that increased secretions are often distressing to the family but typically does not cause discomfort for the patient.

Breathing changes

A change in the breathing pattern is common in patients with DIPG. Oxygen provided by nasal cannula may be beneficial to provide comfort but may also be uncomfortable for some patients. Focusing on what makes the individual patient comfortable is most important. At the final stages of life, the patient will have irregular breathing and will have long pauses in breathing prior to death. Medications that are provided for comfort can ease the breathing pattern and decrease the feeling of breathlessness experienced by the patient. The increased work of breathing and abnormal breathing pattern can be distressing to patients and families and the inclusion of comfort measures/medications may be essential. Examples include opioids and anxiolytic medications.

Posturing (Decorticate/decerebrate)

May appear at the time of brain stem herniation resulting in cardiopulmonary arrest. This is very distressing to the families, yet it important to relay to family, the patient is not feeling pain. Lorazepam or muscle relaxants may help.

Logistics after death

If the patient is in the hospital at the time of death, the medical team will confirm the death, remove equipment and take away medications. Some families wish to help the medical team bathe the body. The medical team will help contact the funeral home at the request of the family.

If the patient is in the home with hospice support, a hospice nurse will come to the home to confirm the death. The hospice staff will help the family bathe the body and contact the funeral home. The funeral home will come to the house and transport the body to the funeral home. The staff will provide support to the family. In the event that hospice is not available for the family, some physicians offer to come to the home for confirmation of death.

Tumor Donation

Families and patients may elect to donate their child's brain and tumor to research through a limited autopsy. Some DIPG researchers have advocated that patients and families be approached more regularly to consider postmortem brain or tumor donation. One recent institutional series demonstrated that approximately half of those approached for autopsy, agreed to the autopsy and tissue donation; no families reported additional distress from the autopsy; and families generally reported deriving comfort from the hope that donated tissue could contribute to research. The study also demonstrated that tissue adequate for analysis was obtained as long as 48 hours after death.

References:

Velduijzen SEM et al. *Palliative and end-of-life care for children with diffuse intrinsic pontine glioma: results from a London cohort study and international survey; 2016; Neuro-Oncology*18 (4).