

## **Satisfactory example of a case study**

Palliative Care in diffuse intrinsic  
pontine glioma of childhood: Challenge  
and Reflection

Word Count: 3496

## Introduction

The philosophy of palliative care provided to children and their families has a number of similarities with the care provided to adult patients. The focus of care remains on maximising the quality of life with a life-limiting illness. This includes symptom management, developing rapport with the child and family, and aspects related to location of care. Examples of differences include careful titration of doses based on weight of the child, the need for both child and family-centred care, variance in tissue donation practices, communication complexities and the myriad of intense emotions experienced by grieving parents.

This case study aims to identify the issues and challenges in paediatric palliative care relevant to a patient with a diagnosis of diffuse intrinsic pontine glioma (DIPG) in the context of complex family dynamics. This family's narrative highlights issues related to timing of referral to palliative care, the use of complementary and alternative therapies (CAM) and, family coping in the face of a rare and life threatening diagnoses. Other issues include tissue donation and approach to sensitive communication with children about their diagnosis and prognosis. Review of both treatment-directed and palliative therapies used will be undertaken, as well as an assessment of personal learning.

## Case history

■■■■ was a child without any significant past history who presented as a 5 year old girl with a twenty-four hour history of diplopia on 7<sup>th</sup> May 2012. The local paediatric service referred her to an ophthalmologist. Review found right sided 6<sup>th</sup> nerve palsy. Subsequently the local paediatrics service performed a computed tomography of her brain (CT) and found a 2.5cm brain stem lesion.

■■■■ was then concurrently referred to ■■■■■ Hospital neurosurgical team, and the oncology service. By the time of these reviews, her left 6<sup>th</sup> nerve palsy had progressed further and she also had horizontal nystagmus intermittently. S.B. at this time reported no headache but had developed intermittent vomiting occurring once per day. The magnetic resonance imaging (MRI) findings were consistent with a diffuse intrinsic pontine glioma (see figure 1), with a lesion involving the entire pons, a compressed fourth ventricle, with no other intracranial lesions detected.

Her family were advised of a survival time of around three month with no treatment, which was offered as an option. The other option discussed was to have palliative radiotherapy which would offer an improvement of survival to around nine months. Radiotherapy could potentially cause short term symptoms however, and also longer term sequelae, such as infertility and cognitive effects, for survivors. It would also mean significant time in the hospital out of the limited time she was likely to survive.

Her parents agreed to radiotherapy and this commenced the next week, by which time she developed unsteadiness, headache and increase in vomiting. She received ondansetron 4mg every 8 hours and paracetamol 500mg every 4 hours as required initially, and then with some progression of symptoms commenced dexamethasone 4mg twice daily, and ranitidine 75mg twice daily for gastric protection. After a six week radiotherapy course she was transiently able to cease the dexamethasone, when improvement was seen clinically and on progress MRI head. Unfortunately, she subsequently had acute onset unilateral weakness and seventh cranial nerve palsy resulting in reinstatement of the regular dexamethasone dose of 4 mg twice day. This was required for a few months, and then weaning was initiated.

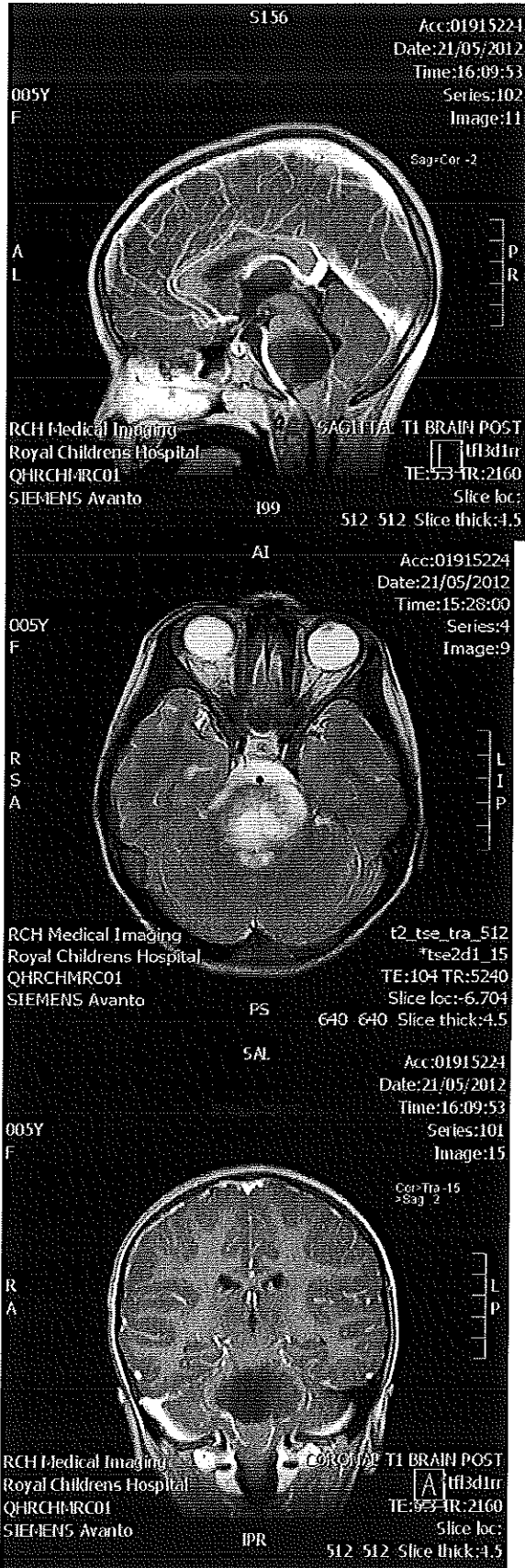


Figure 1: MRI images of the lesion found in S.B. - Sagittal, transverse and coronal views

experienced significant side effects from the ongoing long-term dexamethasone. She had been 19.8kg at diagnosis, and at last weigh was 34.9kg (see figure 2, growth chart). The significant weight gain, in combination with her general deterioration and weakness, affected her mobility. She also experienced cushingoid aesthetic changes and significant striae. The weight gain was actively treated by the family with dietetics support. Later her family instituted a 'blood-type' based diet, after recommendation from a church friend.

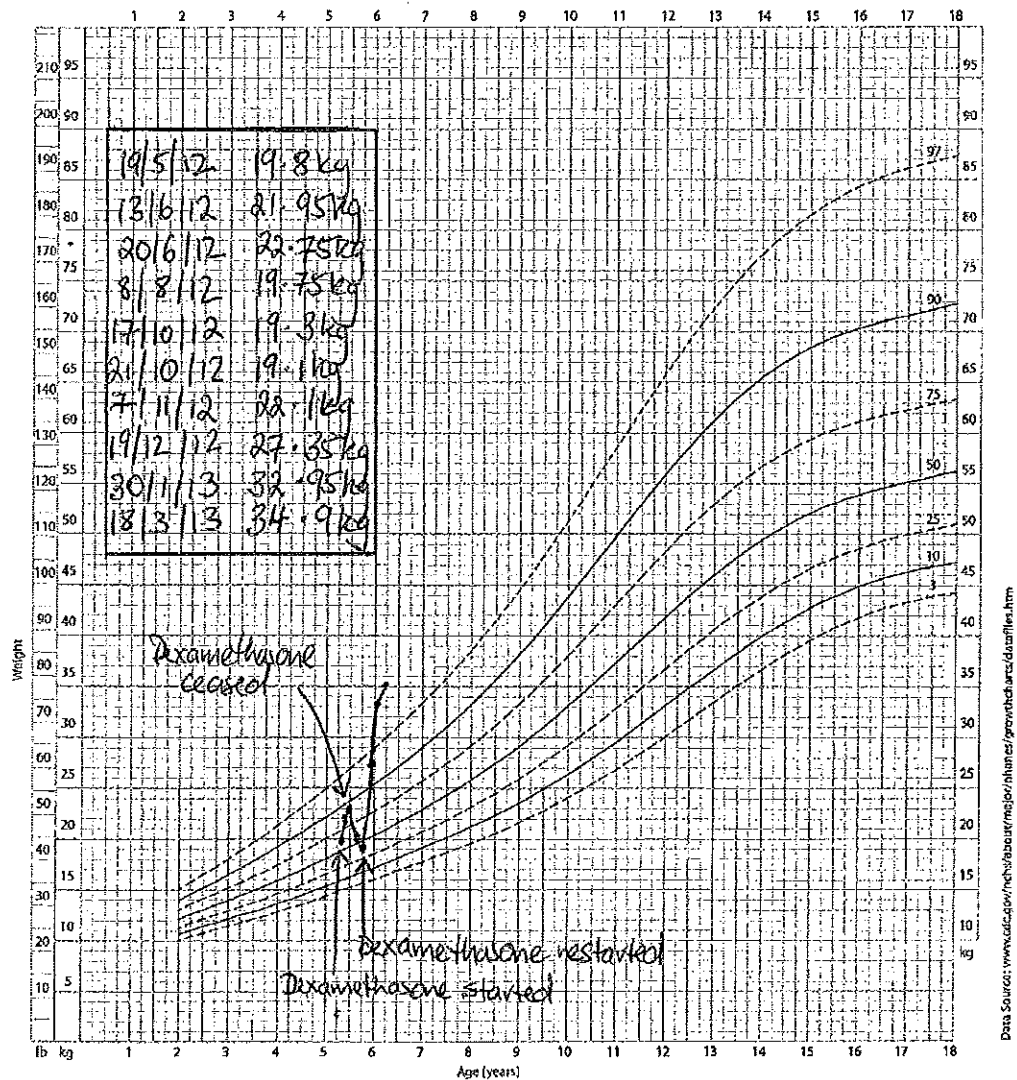


Figure 2 – S.B. Growth chart from diagnosis until death

Inevitable disease progression and hydrocephalus occurred, seen in an MRI in February 2013, coinciding with return of headaches and vomiting. Acetazolamide 250mg twice daily was started, for symptoms of raised intracranial pressure. The dexamethasone dose was also increased from a weaned dose of 1mg twice daily to 2mg twice daily. Oxycodone 3mg every 4 hours as required was prescribed for acute pain episodes. Co-trimoxazole 80/400mg twice daily three days per week was also commenced as Pneumocystis prophylaxis given the long-term steroid use. It was around this time, late January 2013, a referral to palliative care was made by the oncology service. The initial request related to assistance with equipment and modifications at home.

## Social History

■■■■ was the only child of ■■■■ and ■■■■. ■■■■ was previously a teacher but had not worked for some time. ■■■■ had been engaged in studies on accounting when ■■■■ was well, and financially they had been supported on Austudy and received a parenting payment. ■■■■ abandoned his study around nine months into her diagnosis and instead engaged in low income internet-based work. There were significant financial difficulties for this family, and they struggled with petrol costs, parking fees, and maintaining easy contact with the team. Social work assisted in accessing extra funds and the free 1800 number was available to contact the palliative care service.

■■■■ was a Filipino immigrant; ■■■■ and ■■■■ were born in Australia. When ■■■■ was well they had lived in a rural area with a Rural, Remote and Metropolitan Area (RRMA) level 3 classification (1). Initially they were able to stay with friends in Brisbane, and then the family elected to move to the metropolitan area for ongoing treatment. This move decreased personal supports and meant the family needed to access a new local church. ■■■■ found setting up a Facebook page for ■■■■ to be helpful in making new supportive contacts with other families, particularly paediatric oncology families. Their new church was initially very helpful in providing fundraising and spiritual support, however in the later phases G.B. reported feeling pressured by the church to attend regularly even as it was becoming very difficult to mobilise S.B.

During treatment the family accessed several supports for 'memory-making' activities. These included a weekend at the beach through Paradise Kids, a Camp Quality trip to SeaWorld, and a trip to Sydney through the Make-a-Wish Foundation, particularly as ■■■■ wished to go to Taronga Zoo. All of these experiences were viewed positively by the family. Her parents had opted not to directly inform ■■■■ that she would die from her illness, but with the progression of disease and with minimal active treatment, with S.B. herself questioning when she might return to ballet, it's hard to think this bright young lady was not aware.

Despite increasing difficulties with mobilisation, ■■■■ attended school until quite late into her illness. Her school was incredibly supportive, with her class making efforts to work around her limitations, particularly allowing her mother to stay during the day to assist with cares. This was particularly remarkable given her recent transition to the school with the family move, but a school visit facilitated by the oncology and palliative care services may have helped.

## Palliative Care Team Involvement

■■■■ and family were first seen by the Paediatric Palliative Care Service at ■■■■ Hospital in February 2013. Her symptoms at this time included persistent left-sided sixth and seventh cranial nerve palsies, residual left sided weakness, headaches that were well controlled on dexamethasone 1mg twice daily, and acetazolamide, 250mg twice daily, with paracetamol 500mg every 4 hours as required.

■■■■ symptoms were well managed with oral medications. Over the course of five home visits metoclopramide 5 mg every 6 hours as required was added to the symptom management plan in case ondansetron was not effective in relieving nausea. Movicol (macrogol) 1 sachet daily as required was prescribed in anticipation of constipation becoming a problem due to her use of opioids and immobility. Aside from ondansetron her parents did not tend to regularly access any 'as needed' medications. ■■■■ rarely complained of pain, so we respected her parent's assessment of her levels of pain

On 2<sup>nd</sup> May 2013 at a home visit, she had a decreased level of consciousness and was no longer capable of oral intake or medications. The team advised that she was reaching terminal

phase of care and any important visitations or plans should occur immediately. At this time a 24 hour subcutaneous infusion was commenced comprised of 10mg morphine for pain, 10mg midazolam for potential agitation, 2mg dexamethasone to avoid Addisonian crisis as well as assisting with headaches, 10mg metoclopramide for nausea, and 400mcg hyoscine hydrobromide for 'noisy' breathing. She also was charted breakthrough subcutaneous doses of morphine 2mg, midazolam 2mg and hyoscine hydrobromide 200mcg. Her parents did not feel capable of maintaining the infusion pump cares, or providing subcutaneous bolus medications. Fortunately, a paternal aunt arrived at the house willing to take on this responsibility. The doses of morphine and midazolam used (approx 10 mcg / kg / hour) were very modest and were used with the intention of analgesia and anxiolysis, rather than sedation.

■■■■ died an hour after the palliative care team left, just less than twelve months from the beginning of her illness.

## Discussion

### *DIPG diagnosis and management in children*

Overall childhood cancer continues to be a rare diagnosis, but represents the second commonest cause of death in Australia in children ages 1 to 14 years old, second only to trauma. The overall annual rate of childhood cancer in Australia is 157 per million, one-fifth being central nervous system (CNS) tumours (2) and of these around one-fifth are brainstem gliomas. The most common brainstem glioma subtype is DIPG (3).

DIPG of childhood is a devastating diagnosis. DIPG has consistently had a median survival time of 9 to 12 months despite multiple clinical trials of treatment modalities (4). Long term survival for this condition is less than 10% (5). The onset of symptoms prior to diagnosis is primarily over a short period of time, with a median of 4 weeks (range 1-12 weeks) symptom duration and median age at diagnosis is 6.8 years old (range 0.2-14.7). The most common presenting features are cranial nerve palsies (seen in 90%), ataxia (72%) and long tract deficits (54%) (4). Diagnosis occurs through magnetic resonance imaging where an infiltrative lesion occupying two-thirds or more of the pons is seen. The MRI appearance is so characteristic that a tissue biopsy is not warranted in most cases (3).

As per the child in this case study, the primary treatment modality currently is radiotherapy. Around half the patients in selected facilities are now also receiving chemotherapy as a part of an ongoing clinical trial (4). The likely vascularity of the lesion has led to some work into endothelial growth factor inhibiting agents, such as vandetanib. Although survival gains found experimentally have been small (5), this therapy may have an important steroid sparing effect, and reducing high burden of steroid side effects may improve quality of life. Surgical resection is not an option in DIPG related to the infiltrative nature of the lesion as well as the actual site of the lesion (3). Ongoing work into other methods of management are key in both improving survival and avoiding dexamethasone, given its high burden of side effect, particularly seen in this case.

### *Effects of Dexamethasone*

Initial targeted therapy of dexamethasone at tumour swelling lead to the use in end-of-life for ongoing symptom management for this patient. In cases of high-grade glioma it may cause symptomatic relief by reducing peritumoral water content and surrounding tissue pressure, thus possibly increasing perfusion to oedematous brain tissue (6). Around 50% of adult-patient neuro-oncologists routinely use dexamethasone, at the dose ■■■■ was given, and then attempt to wean post-radiotherapy. The most common side effects on chart and survey review have been found to be weight gain, insomnia, proximal muscle weakness, and gastrointestinal symptoms in decreasing frequency (7). Unfortunately the resulting sequelae for S.B. were significant and distressing for the family, including weight gain and mobility restriction,

striae, self-esteem and comfort issues. Dexamethasone was very effective in this case in controlling the headache symptoms, however it was vital to continue to consider if the side effects were worth the improvement in symptoms.

The restriction in mobility was significant for both the patient and the family. Her parents were having difficulties in caring for her, particularly transferring her including for bathing. The occupational therapist from the palliative care service reviewed the home and made bath seat recommendations. However the small size of the house and particularly the bathroom made the use of a hoist impossible. ■■■■■ did develop some significant back pain related to cares for her daughter.

The striae were quite significant and she experienced friction between affected areas of weakened skin when mobilising, with associated pain. There were areas of epidermal tearing up to a centimetre wide around her axillary area, upper thighs and abdomen. This issue was partly addressed with duoderm patches to provide strength to the skin and avoid the friction element of the discomfort.

On many of the home visits ■■■■■ would show the palliative care team photos and videos of the daughter 'she used to have' with ■■■■■ in the room. This was concerning as ■■■■■ was perfectly capable of understanding the conversation, and may have added to issues with self-esteem she already had about 'being different', gaining weight, and becoming increasingly dependent.

#### *Blood-type diet*

The blood type diet adopted in this case was developed by Dr Peter D'Adamo, an American 'Naturopathic Physician'. Although his diet system has largely been discredited in scientific experimentation, it continues to have a wide popular literature base (8). The blood type diet is based on the idea that lectin proteins in food have a chemical reaction with the blood antigens, primarily the ABO antigens. The reaction leads to lectins 'agglutinating' in organ systems, such that modifying diet to suit your genetically inherited blood type promotes well being. With ■■■■■ having an A type blood group she was recommended a vegetarian diet with a range of vegetables, grains and fruit, but was to avoid dairy and wheat (9). The blood-type diet involved a good range of healthy foods but in the terminal phase it did appear unnecessarily restrictive for ■■■■■ who wanted chocolate, as well as greater amounts of food overall. The institution of this diet did not lead to any perceptible weight loss. The use of diets such as this, and other CAM, demonstrates the family's desire to explore all options in the hope that their child may get better.

#### *Integration of Palliative Care in Oncology Treatment*

Referral to palliative care occurred in this case when the treating oncologist needed extra resources and assistance for home care provision. An argument though could have been made for much earlier referral, perhaps even at diagnosis, given the known poor prognosis of diffuse intrinsic pontine glioma. This early introduction could allow for better establishment of rapport and time for the parents to have a better understanding of the breadth of paediatric palliative care. While it may be considered that oncologists have independent skills in palliative care and that avoiding 'abandonment' of their patients at illness progression is important, early referral may assist in patients viewing the two disciplines as 'on the same team' with a gradual transition process.

Early integrated referral has been recommended in a number of reviews (10). The barriers in referral include misconceptions from health professionals (11). This reluctance for referral is also found to be exacerbated in hospital settings, and in more senior doctors (12). This reflects a dual role necessary for palliative care staff, the integrated multidisciplinary clinical care of children with life limiting illness, and the need to educate colleagues and the

community about the true benefits of palliative care referral. Benefits include better advance care planning, fewer deaths occurring in intensive care, less suffering for children, and better preparation for end-of-life care (13). In adults it has even been demonstrated that earlier referral can lead to longer life expectancy (14).

#### *Limitation of active treatment*

This patient did not have formal documentation of what cares would be appropriate in the event of deterioration. Factors in this were difficulty of the parents facing this eventuality, and also partly as it would not be necessary if end-of-life cares were maintained at home without ambulance involvement. One discussion that did occur was at the last home visit from palliative care, around the issue of [REDACTED] no longer being able to tolerate oral intake. Her parents assumed ongoing nutrition would be required, however it was discussed that this was likely to be unhelpful in the face of a short survival time, and could lead to vomiting and increased symptomatology. The parents were able to reflect on this and agreed not to pursue nutrition, and this discussion may have helped the parents see that terminal phase had been reached.

#### *Organ Donation and Tissue Donation for Research*

In this case the family did not seek tissue donation, and the only direct questioning from health professionals was regarding tumour tissue donation. Although the family agreed in practice to the idea of donating the tumour, they could never bring themselves to fill in the paperwork, and she died before these were ever completed. Other tissues that could have potentially been donated in a non-metastatic solid tumour case like this include corneas, heart valves, skin and bone fragments.

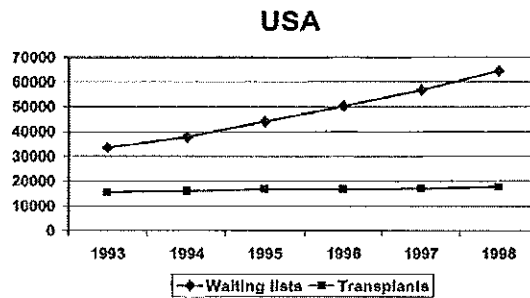
This 'opt-in' plan is a common stance of the paediatric palliative care paradigm, due to factors including effects on rapport, distress caused to parents, and effects on the perception of palliative care by both other health-care workers and general society. This has to be balanced with the paucity of viable organs available to the community for transplantation, which shows a trend overall towards increasing demand without increasing supply (see figure from 15). Particularly for DIPG, poor sample availability is blamed for lack of progress into alternative treatment options (5). The other consideration is later regret of the parents on having missed out on this option, as was repeatedly reported in a study on grieving parents (16). Perhaps this may be related to some feelings in parents of having missed finding 'meaning' in the devastating loss of a child.

#### *Communicating with children about their prognosis*

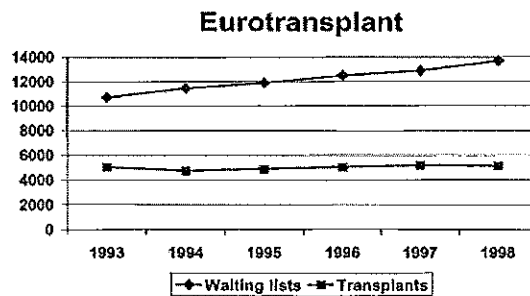
The patient had been an avid ballet dancer prior to her illness, and would ask about when she would return to dancing. This, in a way, asked a bigger question of what was happening to her, and where was all of this going. The parents in this case avoided directly answering the question, and told her to focus on taking her medications and praying to God. Other options may have been to reflect back to the child what the child felt was happening, or to use the question as a doorway to this bigger discussion. While the parents in any case remain the expert on their child, and the appropriate judge of what should be discussed and how, there are many benefits to open discussion that have been identified. Benefits seen have included reduced anxiety, depression and behavioural problems in children (17), and a significant proportion of parents retrospectively experience regret at not having had this discussion (18). Of course, it is also critical also to consider the developmental level of the child as highly relevant to tailoring a discussion.



## The Widening Gap 1993-1998



Source: UNOS



Source: Eurotransplant

FIGURE 1. Difference between organ supply and demand in the US and Eurotransplant (Austria, Belgium, Luxembourg, Germany, The Netherlands) in 1997.

Figure 2 – The increasing demand for transplants

### Personal Reflection

I was interested to find out about the social media links that existed between the oncology families. Although in this case █████ seemed to find the Facebook links helpful, perhaps given a degree of isolation with the recent move, I was not sure if these links were always a positive experience. Having worked with community-based cancer support previously, I had seen friendships develop between oncology patients, and then the impact of subsequent deaths. This seemed similar in some ways, with █████ asking us about other families known to our service she had connected with on Facebook, and seeing children with the same diagnosis pass away. There was one particularly graphic video uploaded by a family, showing them carrying their child's body from the house.

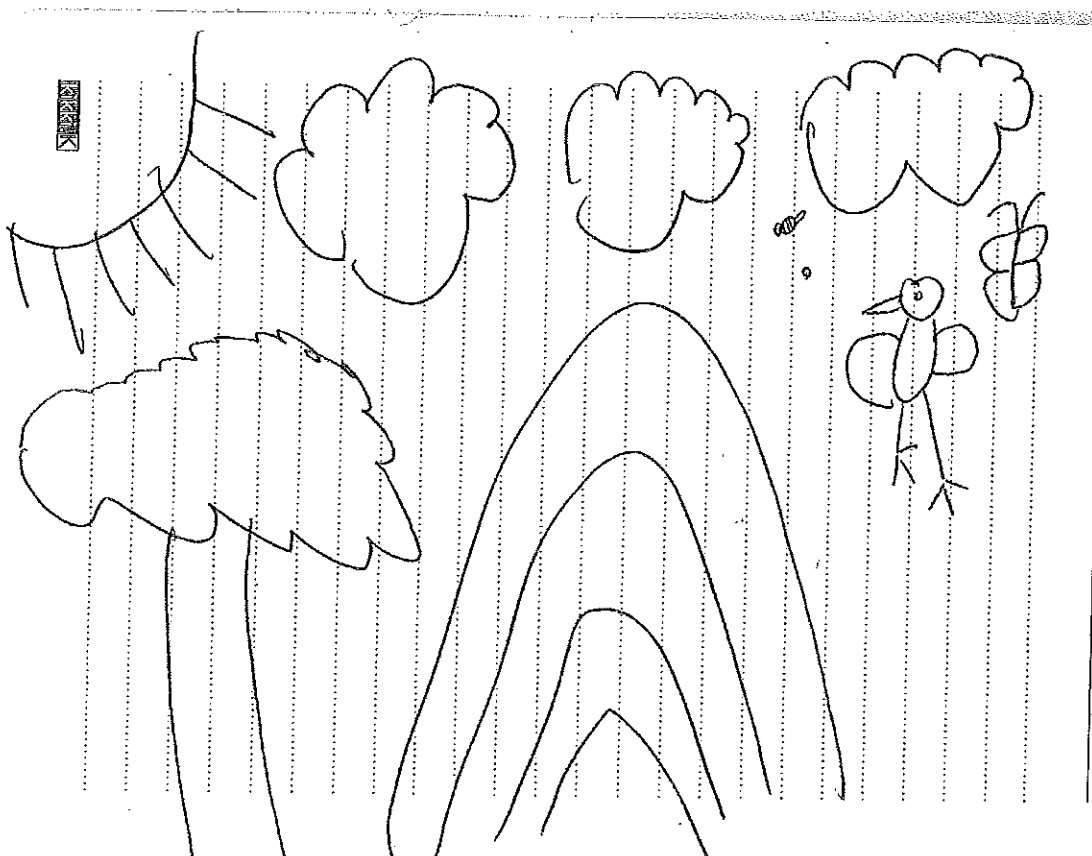
During the time we had the privilege to be a part of █████ life; I grew very fond of her. When she passed away I was not able to attend her funeral due to timing constraints. The ethics and appropriateness of whether to attend or not is controversial. In the circumstances of having frequently seen them for long periods during this intense time it may have been appropriate. It was a comfort to know that a card was sent from the service, and bereavement follow-up would be offered. I found it invaluable to attend to self-care at this time, and was well supported in this by the palliative care team.

Rapport is critical in palliative care, and I felt that the family engaged well with us. We took time to hear and validate their fears and support their goals, as well as taking time to get to know █████ herself. It was challenging at times not to let judgement impede this rapport, when we disagreed with some of their choices such as the dietary restrictions, lack of accessing 'as required' medications and choice not to openly discuss disease progression with █████

I had not had a great deal of experience with patients who had died before this term, having worked solely in a developed country with low overall mortality rates. I plan in the future to improve in my ability to discuss painful topics and pre-empt symptomatology relevant to specific diagnoses. This term was invaluable and I remain very grateful to all the families who let me into their lives, and homes, at a vulnerable time, and trusted me with their most personal hopes and fears.

### Conclusion

This case study highlights some of the complexities of paediatric palliative care. This included managing symptoms and supporting the child and parents with a severe disease burden. Considering the family context, including faith and alternative medicine, was key in implementing effective interventions, optimising the limited time left for the patient and maintaining ongoing rapport between the family and health care professionals. There were many lessons to be learnt from this family, including generic principles related to the provision of paediatric palliative care, as well as issues unique for this particular patient. For example, generally, careful communication about the future is important for all children with life limiting conditions. In this case the approach needed to be balanced to the specific needs and desires of [redacted]'s parents. Early integrated referral to palliative care should be promoted where appropriate to facilitate honest communication between family members, to explore wishes related to resuscitation, and determine other goals of the child and family (e.g., preferred location of care and preferences related to tissue donation possibilities).



## References:

1. Australian Institute of Health and Welfare. 2004. Rural, regional and remote health: a guide to remoteness classifications. AIHW cat. no. PHE 53. Canberra: AIHW.
2. Baade, P.D; Youlten, D.R; Valery, P.C; Hassall, T; Ward, L; Green, A.C. and Aitken, J.F. 2010. Trends in incidence of childhood cancer in Australia, 1983-2006. *British Journal of Cancer*, 102: 620-626
3. Grimm, S.A and Chamberlain, M.C. 2013. Brainstem Glioma: A Review. *Current Neurological and Neuroscience Reports*, 13: 346-353.
4. Hargrave, D. and Chuang, N. 2008. Conventional MRI cannot predict survival in childhood diffuse intrinsic pontine glioma. *Journal of Neurooncology* 86: 313-319.
5. Broniscer, A; Baker, J; Tagen, M; Onar-Thomas, A; Gilbertson, R.J; Davidoff, A.M; Panandiker, A.P; Leung, W; Chin, T.K; Stewart, C.F; Kocak, M; Rowland, C; Merchant, T.E; Kaste, S.C and Gajjar, A. 2010. Phase I study of vandetanib during and after radiotherapy in children with diffuse intrinsic pontine glioma. *Journal of Clinical Oncology*, 28(31): 4762-8.
6. Bastin, M.E; Carpenter, T.K; Armitage, P.A; Sinha, S; Wardlaw, J.M and Whittle, I.R. 2006. Perfusion and Water Diffusion in Patients with High-Grade Glioma. *American Journal of Neuroradiology*, February 27: 402-408
7. Sturdza, A; Millar, B.A; Bana, N; Laperriere, N; Pond, G; Wong, R.K. and Bezjak, A. 2008. The use and toxicity of steroids in the management of patients with brain metastases. *Supportive Cancer Care*, 16(9): 1041-8.
8. Eat right for your type. 2012. <http://www.dadamo.com/> [accessed online 23<sup>rd</sup> August 2013].
9. Lam, M. 2013. Blood type diet. [http://www.drlam.com/blood\\_type\\_diet](http://www.drlam.com/blood_type_diet) [accessed online 23<sup>rd</sup> August 2013].
10. American Academy of Pediatrics. Committee on Bioethics and Committee on Hospital Care. 2000. Palliative care for children. *Pediatrics* 2000; 106: 351-357.
11. Monterosso, L and Kristjanson, L. 2008. Supportive and palliative care needs of families of children who die from cancer: an Australian study. *Palliative Medicine*, 22(1): 59-69.
12. Kogan, A.C; Brumley, R; Wilber, K and Enguidanos, S. 2012. Physician Factors That Influence Patient Referrals to End-of-Life Care. *American Journal of Managed Care*, 18(11):e416-e422.
13. Wolfe, J. 2011. Recognizing a Global Need for High Quality Pediatric Palliative Care. *Pediatric Blood and Cancer*, 57(2): 187-188.
14. Temel, J.S; Greer, J.A; Muzikansky, A; Gallagher, E.R; Admane, S; Jackson, V.A; Dahlin, C.M; Blinderman, C.D; Jacobsen, J; Pirl, W.F; Billings, J.A and Lynch, T.J. 2010. Early Palliative Care for Patients with Metastatic Non-Small-Cell Lung Cancer. *New England Journal of Medicine*, 363:733-742.
15. Cohen, B. and Wight, C. 1999. A European Perspective on Organ Procurement: Breaking down the barriers to organ donation. *Transplantation* 68(7): 985-990
16. Oliver, R.C; Sturtevant, J.P; Scheetz, J.P. And Fallat, M.E. 2001. Beneficial Effects of a Hospital Bereavement Intervention Program after Traumatic Childhood Death. *Journal of Trauma-Injury Infection & Critical Care*, 50(3): 440-448.
17. Sutter, C. and Reid, T. 2012. How do we talk to the children? Child life consultation to support the children of seriously ill adult inpatients. *Journal of Palliative Medicine*, 15(12): 1362-8.
18. Kreicbergs, U; Valdimarsdottir, U; Onelov, E; Henter, J. and Steineck, G. 2004. Talking about Death with Children Who Have Severe Malignant Disease. *New England Journal of Medicine*, 351(12): 1175-1186.

Dedicated to [REDACTED]

*A wonderful family who taught me more than  
they will ever know*

