

# A Case of Anencephaly: Integrated Palliative Care

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## ABSTRACT

Following a diagnosis of anencephaly at the 24 week anatomy follow-up scan this case study exhibits an approach that integrates the primary, secondary and community services while maintaining the family unit at the centre of care, when an outcome to a pregnancy is not what was originally envisaged. Hope's journey illustrates how integration of the multidisciplinary hospital team and community care can assist and support the family when planning a way forward, tailored to the family's personal, physical, emotional and spiritual needs. After her mother's full term pregnancy and vaginal birth after caesarean section (VBAC), Hope was discharged home seven hours after her birth as her parents wished for palliative care. She lived 14 hours, a life filled with love, dying in her family home as her parents wished. This pregnancy and outcome was a highly emotional journey. The care the family received was compared to that of a perinatal palliative care model and identifies strategies that could be adopted by maternity teams for similar cases in the future.

## INTRODUCTION

Diagnosis of a fetal condition in pregnancy that is incompatible with life, such as anencephaly, presents a challenge for practitioners. Families faced with these situations need unbiased, sensitive and complete information to enable them to make informed decisions and plan for their pregnancy, labour and birth care.

This case study describes the journey for a family when they decided to continue the pregnancy and build a support network to meet their personal, physical, emotional, cultural, religious and spiritual needs. The parents have given consent for the author to tell their story and to use the photograph of Hope. For this story the parents will be referred to as Sally and Harry.

The decisions made, the planned care pathways, and the outcomes achieved, are described. Possible frameworks of care and improvements for future cases are described.

## ANENCEPHALY

Anencephaly occurs in around 1 in 10,000 pregnancies and is a neural tube defect that results from failure of the neural tube to close around

25-27 days following conception. This leaves the developing forebrain and brain stem exposed. The forebrain is either destroyed or fails to develop. This means the cerebrum and cerebellum are reduced or absent, but the brain stem is present (Blackburn, 2003). If the babies are born alive, they are most likely to live only for a few hours and at the most no longer than 2 weeks. Anencephaly is usually diagnosed by maternal serum screening (raised serum alpha-fetoprotein) and/or ultrasonography and usually results in a termination of pregnancy, stillbirth or early neonatal death (Tadanori & Hideki, 2013).

## BACKGROUND

Sally is a 32 year old mother of two, married and in a stable, well supported relationship. Her first child, a boy, was a normal birth at term, and weighed 3325gms. A daughter was born later that same year by elective Caesarean section. Sally felt there had been too short a time since her last birth and did not feel resilient enough to proceed with a vaginal birth following the third degree tear she sustained during her first birth. Sally had experienced miscarriages in 2006, 2010 and 2011, is a non-smoker and has a Body Mass Index (BMI) of 23. During her pregnancy, Sally took Sertraline tablets, 50mg daily, because of her history of postnatal depression. There is no other significant family or medical history.

When pregnant with Hope, Sally had a 20 week anatomy scan which stated "poor views of the fetal skull" and a repeat scan was suggested at 24 weeks. This subsequently diagnosed anencephaly. The World Health Organization (WHO), (2013), suggest where the fetus has anencephaly that early identification and ongoing assessment, which includes the family, reduces suffering within the family structure and this is where the palliative care pathway begins in this case.

## PALLIATIVE CARE PATHWAYS

In 20-40% of pregnancies where fetal abnormality is identified as being incompatible with life, parents choose to continue with the pregnancy rather than agreeing to termination (Engelder, Davies, Zellinger & Rutledge, 2012). Perinatal palliative care is an option for parents who wish to continue with the pregnancy. It commences at antenatal diagnosis, or prior to discharge from hospital. The National Association of Neonatal Nurses (NANN) position statement on palliative care for newborns and infants focuses on maximizing the quality of end-of-life care to support a "Peaceful, dignified death for the infant and the provision of loving support to the family and healthcare providers" (NANN, 2010, p. 287).

The British Association of Perinatal Medicine (2010) framework covers routine pre- and postnatal birth planning to include survival, or end-of-life by natural causes, and post end-of-life care. The framework covers family care, communication, documentation and the importance of flexible parallel care planning. Additionally, the Association for Children's Palliative Care (ACT, 2009) describes a neonatal pathway for babies needing palliative care with the aim of ensuring that appropriate care is given at the right place, at the right time. It consists of three stages and is guided by six standards to ensure the baby and the family is always

positioned at the core of the care planning process. This was the pathway chosen in the case described here.

## STAGE ONE: ENTRY TO THE PALLIATIVE CARE PATHWAY (ACT, 2009)

### FIRST STANDARD OF BREAKING THE NEWS:

Breaking the news to the family (including siblings) may require emotional, religious and spiritual support. Discussion needs to cover future baby care needs, preparation for the place of birth, and identification of people who will be involved in future care e.g. General practitioner (GP), midwife, hospital team, social worker and information about legal requirements for registering the birth.

In this case the ultrasonographer informed the parents that their baby was anencephalic at the 24 week ultrasound scan. This diagnosis was followed by a visit to the private obstetrician. However, this relationship broke down as the obstetrician was not prepared *“to journey with us in the way that we wanted”* (parents of baby). Thus they felt unsupported to continue the pregnancy under the obstetrician's care. This highlights Sumner, Kavanaugh & Moro's (2006) recommendation that parents need an environment where they can experience open communication and information about care options.

Leuthner (2004) believes there are three important discussions to have with parents when breaking devastating news. These are: certainty of diagnosis, prognosis, and the meaning of the prognosis for the family. This latter includes describing to parents how their baby will look and act (Engelder et al., 2012). For Sally and Harry, a follow-up appointment was made to see a visiting Maternal Fetal Medicine (MFM) specialist who reconfirmed the diagnosis and explained to the parents it was unlikely their baby would survive birth, or live for very long after birth. He also discussed the choice of continuing or terminating the pregnancy and the option of VBAC to assist an earlier recovery for Sally. Once the decision to enter the palliative care pathway was made, then the plans for the birth, and the anticipation of both a birth and a loss could begin. This included hellos and goodbyes in the grief process as suggested by Sumner et al. (2006).

Sally and Harry returned to their GP for advice, information and choices in care. They required a team that would listen and respect their decision making and personal needs during this special and sensitive chapter of their life. Their GP accessed a midwife Lead Maternity Carer (LMC) to provide care in partnership and, most importantly, to support the parents' wishes to give birth to their baby, and to take her home should she survive. An obstetrician who was willing to support them was also nominated in case complications developed. Discussion took place as recommended by the referral guidelines (MOH 2012) and it was agreed it was appropriate for the midwife to continue as LMC in order to achieve the requested outcomes even though in reality the care would be shared. Together the midwife and parents developed a birth plan which involved discussion, open communication and documentation in the clinical records. This fulfilled the second standard of the palliative care pathway of *“Planning for going home”*. Below Sally describes the care she received during this waiting period.

*I was offered endless help, meals, childcare, and given gifts. Hope was given gifts and people respected our decision and looked forward to Hope's arrival. She was a very much wanted baby not just by us but also by our extended family “our village”! So many lovely things were coming our way, cards, flowers, well wishes and warm words of encouragement; I decided to start a journal to record such acts. By the time of Hope's expected due date, there were over 100 items listed in my 'Book of Hope'.*

Pictures that tell the baby's story remind parents how much their baby was loved, and how their life had a purpose (Rosenbaum, Smith &

Zollfrank, 2011). Similarly, Sumner et al. (2006) found that gathering keepsakes, along with the experience of compassionate quality care, stays in the memory of families forever. Further, bereaved parents with positive memories were more likely to have experienced effective palliative care according to Rooy, Aladangady & Aidoo (2012).

### DECISION MAKING

Harry and Sally felt it essential that the health professionals caring for them maintained their family at the centre of care but also attended to their personal emotional, spiritual, cultural, and physical needs. They requested support to continue their pregnancy; they wanted a plan in case of misdiagnosis; and a plan for neonatal palliative care with a wish to take their baby home if she were to survive labour. ACT (2009) identifies that around 90% of neonatal deaths occur in the clinical hospital environment and recommends that some of the babies, especially the ones with no cure, be transferred to their own home or a children's hospice with appropriate support. This small District Health Board (DHB) does not have a designated children's hospice or perinatal palliative care policy but it can tailor care to the individual's needs, which happened in this case. Sumner et al., (2006) discuss the importance of caring for the family and baby with dignity and respect and recommend that integrated palliative care programs be introduced into institutions. Following this event the DHB is reviewing its palliative care policy with the intention to include perinatal palliative care in the new policy document.

## STAGE TWO: LIVING WITH A LIFE THREATENING OR LIFE LIMITING CONDITION (ACT, 2009)

Sally and Harry were both aware of the probable outcome of stillbirth or early neonatal death for their baby so vaginal birth was chosen rather than a repeat Caesarean section because this would allow them to spend more precious hours with their baby if she survived. It would also increase the likelihood of taking her home alive in the hours following her birth. They were fully informed and aware of complications that may be a consequence of having an anencephalic baby such as polyhydramnios, prolonged pregnancy and stillbirth, but they did not want to discuss a plan for a funeral for their baby. Sally said,

*We decided we would continue with the pregnancy, she was ours and we would not give up on her existence despite the obvious painful journey ahead. The odds were not in our favour; however we decided to face the situation with courage and love. We focused on bringing our little girl home alive, she would (in our minds) survive birth, breastfeed, be held by loving friends and family and her life would be celebrated. She was chosen for us and we for her. We would give her a birthday and not plan for a funeral.*



## COMMUNICATION

Verbal and written communication with Sally and Harry, the GP, obstetrician and midwife allowed the professionals to understand the parents' wishes which were to ensure *"a seamless and united approach to my care"* (Mother of baby). Even though they did not want to discuss the death of their baby, Sally and Harry were fully aware this was going to be what they faced at some stage. They made it clear they wanted to address the positive aspects of this pregnancy and birth by having a plan in place for all eventualities. They declined social work referral but their situation was discussed and documented in the minutes of the monthly complex care meeting which the maternal social worker, obstetricians (including the visiting MFM specialist) and neonatal team attend. The multidisciplinary team was thus fully aware of the parents' wishes and plan of care. This multi-agency assessment of the family's needs and care plan addresses stage two of the perinatal palliative care pathway (ACT 2009).

Sally and Harry agreed to make a birth plan but did not want to meet personally with the neonatal or paediatric team. They were happy for this communication to be carried out by the midwife (LMC). A written referral was sent to the neonatal team to inform them of the impending birth and the wish of the parents to take their baby home alive should vaginal birth be achieved. To do this it was necessary to be listened to and have a plan for palliative care and support in place. The neonatal team responded by reassuring the midwife, GP and parents that they were aware of the upcoming birth and they were happy to meet the parents and discuss their needs after the birth, if desired.

Discussion surrounding the birth plan included questions such as: what happens if Sally has a prolonged pregnancy? What happens if labour does not commence spontaneously? Would she continue with the pregnancy and further the risk of stillbirth or would she choose the risks of elective caesarean section over the risks of induction of labour? Induction methods such as stretch and sweep, homeopathy or cervical balloon ripening techniques would be offered as opposed to pharmacological induction using prostaglandins or oxytocin. This was in view of Sally's previous Caesarean section. The potential advantages and risks were discussed for Sally and her baby.

Auscultation of the fetal heart in labour was also discussed. In view of the previous caesarean section continuous cardiotocography (CTG) would normally be recommended. However, there was no advantage given that her baby was expected to die in labour or shortly after the birth. Should the fetal heart be recorded what would happen if abnormal patterns were detected? Would the parents want to listen to the fetal heart to know whether their baby was still alive in labour? The parents were unsure, so the birth plan said that they would decide on the day if they wished to hear the fetal heart beat. Sally believed that she was very well tuned in to her baby's movements and did not see auscultation as a priority.

At term plus nine days Sally and Harry attended the hospital for assessment with a view to inducing labour but, during their journey to the hospital, Harry informed Sally that he felt they should wait a little longer and after further discussion this was agreed the best way forward if Sally was still keen for a VBAC. The very next day, at term plus ten days Sally was admitted in spontaneous labour with her support crew. The parents declined the use of the nominated hospital room for labouring couples expecting baby loss. They had personal baby clothes ready to dress baby Hope and the family and the minister were on call in case they were required.

During labour Sally played spiritual music and positive affirmations collected during her pregnancy were displayed on the walls surrounding her. She also had a book of spiritual scripts that could be read if she requested. The atmosphere was relaxed. There were times of laughter and joy, and times of quiet and tears. Sally's labour progressed normally with her using the birth pool for pain relief only. She breathed through

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Further plans would be made on a day by day basis tailored to the family's individual needs.

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the birth to try to avoid a further third degree tear and, four hours after admission, baby "Hope" was born in good condition, covered in vernix. She had good tone, was dusky in colour and attempted to cry. The midwife was unsure how long she would live so Hope was passed to mum and dad for skin to skin contact; normalizing and celebrating the birth being the priority. Sally had a small first degree tear which did not require suturing. The photographer had taken pictures of every detail of the room and captured the moment of birth; photos that are now everlasting memories.

There was a gradual change in Hope's appearance as she began "pinking up" beautifully and mouthing for the breast which was amazing to see. Sally latched her to her breast unassisted and we all observed an amazing breast feed. Siblings and the rest of the family came to meet baby Hope shortly after her birth. They watched Hope being checked, measured, weighed and dressed. Following her feed she was cuddled by all. The paediatrician was informed of Hope's birth and the fact that Sally and Harry planned to stay in the hospital overnight before heading home with Hope the next morning.

## STAGE THREE: END OF LIFE AND BEREAVEMENT (ACT, 2009)

This stage includes the 2 standards of an end of life plan and continuing bereavement support. The paediatrician visited Hope on the Delivery Suite and planned her discharge home with the staff, LMC and family. Assessment of pain and signs of distress were discussed with the parents and paracetamol was prescribed to be administered to Hope if the parents thought there were signs of distress. Kangaroo nursing and comfort measures were also encouraged. Further plans would be made on a day by day basis tailored to the family's individual needs. A neonatal home care nurse would also be available if necessary. Plans were also made for the maternal social worker to visit the family at home to provide social, financial and counseling support. The family church minister was in contact with the family to address their religious and spiritual needs.

During that evening Hope continued to feed at the breast. She passed urine and meconium. However, she did have a mucousy, dusky episode. This event reminded the parents of their intention to return home with baby Hope alive if they could. The core midwives were aware of the family's wishes and assisted the LMC to organize a prompt discharge. Sally and Harry were given the direct telephone numbers for their LMC midwife, the ward and the consultant paediatrician so they could phone or text if they had any concerns overnight. The night staff were also aware of these arrangements.

At the family home Sally's parents and sister as well as her children and husband took turns to rest, love and hold baby Hope. Unexpectedly,



without warning, Hope stopped breathing and died some 14 hours after her birth. A phone call to the hospital ward to inform the staff of the events resulted in the consultant paediatrician driving the half hour to Sally and Harry's home to certify baby Hope as having died at 5.30am.

The community, local church minister, friends and family provided food and support for Hope's service of celebration and remembrance. Some 200 members of the community rallied around the following few days to set up and attend a memorable service to say "good bye" to baby Hope, with a final balloon release before she set out on her last journey.

## CONTINUING BEREAVEMENT SUPPORT

This town has an active Stillbirth and Neonatal Death Society (SANDS) which works closely with the maternity unit. The organization has raised funds to provide 'keepsake' boxes to parents who experience baby loss. These boxes contain patchwork quilting, baby charms and keepsakes, inkless foot/handprint sets, as well as contacts for support from their group, along with suggested readings.

Long term follow-up of families who experience perinatal loss is recommended and may last for up to a year (Engelder et al., 2012). The New Zealand model of maternity care assists with continuity of care within this palliative care model and often LMCs remain in contact with these families beyond the 6 weeks allocated to the postnatal module, providing support rather than care. The benefit of having the GP involved manifested in that on-going treatment and care occurred with the knowledge of the journey the family had been on. Additionally the maternal social worker was available for ongoing support. Interestingly Branchett & Stretton (2012) found that experiences of bereavement care were more positive where many staff participated in the care rather than one specific midwife/nurse. This would seem to support the need for multidisciplinary support for the family and the LMC.

## PARENTAL REFLECTION

When Sally reflected on her care and experience, she said she appreciated the 'team approach' with the multidisciplinary collaboration that took place and the offer of follow up appointments, extra support and information. She felt heard, respected and supported by the professionals involved in her care, as well as by the community that surrounded her in hospital and at home. Having unlimited access to friends and family who visited in hospital, meals supplied, offer of childcare and acts of kindness assisted Sally in her grieving process.

The GP and midwife LMC had both thought about having a discussion in the antenatal period on baby organ donation and having student doctor and student midwife involvement. However, these discussions did not take place with the parents because of their strong desire to take their baby home. A further barrier to these discussions may have been the late involvement of the midwife LMC in the pregnancy, who may have assumed that these discussions had already taken place. However, this has given rise to a question from Sally following the birth. Having had a positive experience, she now, in hindsight, would have liked to have been offered the opportunity for students to learn through her experience, as well as to have had the opportunity to consider organ donation.

Not participating in organ donation may have been a lost opportunity, but would the experience have been the same? Sally suggests that, in the future, if she were to birth another anencephalic baby she would like to explore the option of consenting to organ donation when the baby is about 10 hours of age in order to help the survival of other babies. However, it is recognized as being a sensitive and difficult subject that may not be appropriate to address in all situations or cultures.

## CONCLUSION

The decision for these parents to continue the pregnancy to full term after the diagnosis of anencephaly remains in their opinion the right decision. They feel it has given them comfort in the fact that they had a positive bereavement experience and will not be wondering "what if?" if the decision had been to terminate the pregnancy.

This case illustrates how perinatal palliative care pathways can support and assist professionals working in maternity units when parents decide to continue a pregnancy with a baby with a terminal condition. It also provides a framework to facilitate the parents to have the option of taking their baby home to die with appropriate support in place.

This case study illustrates how the parents can be kept at the centre of care providing space for them to make informed choices with the support of integrated care from both the hospital and community. The key to this family's positive bereavement experience was having an identified person to coordinate care as well as a multidisciplinary support network for the midwife LMC and family. The midwifery partnership and the continuity of care model is the ideal model within which to lead and provide coordination of care for these families and can be achieved with discussion and agreement between the woman, midwife and obstetrician.

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