Parents' submission to London Hospital Clinical Ethics Committee (July 2015)

Our unborn daughter was diagnosed with Trisomy 18 at 23 weeks' gestation.

Traditionally a diagnosis of trisomy 18 has been seen as 'lethal' or 'incompatible with life'. It is certainly a condition associated with significant mortality and morbidity. The majority of fetuses with Trisomy 18 miscarry or are stillborn before term (Morris et al 2008). Of the minority who are born alive, the most recent UK population study suggested that 60% survive at least one week, 39% survive at least one month, 20% at least 3 months and 8% are still alive at a year (Wu et al 2013). Increasing gestational age and female sex are associated with better than average survival rates (Wu et al 2013).

Survivors universally have significant disabilities, including profound developmental delays. Despite the severity of the condition, 1% of children are still alive at age 10 and some people survive into their 20s and 30s.

Perhaps because of the high rate of mortality and the significant disabilities among survivors, it is common in the UK not to offer active treatment to infants with a prenatal diagnosis of Trisomy 18. Instead a palliative care or comfort care approach is offered. However there has been an increasing movement in some countries for parents to opt for and to be offered a more active approach to management of their babies with Trisomy 18. Evidence suggests this may prolong life, with increased survival to a year and beyond for infants offered active treatments (Kosho et al 2006, Bruns 2010).

When we considered the care we wanted for our daughter if she is born alive, we considered:

a. Whether an active approach to treatment would prolong her life. We define 'active approach to treatment' as not withholding any treatment merely on the basis of a Trisomy 18 diagnosis; instead treating according to the baby's clinical condition. In our daughters case, this may include surgical repair of diaphragmatic hernia/eventration and cardiac defects.

b. Whether the treatment in itself would cause our baby pain and suffering, and if so, where the balance lay between the benefits of the treatment in terms of prolonged life and the burdens of active treatment for her.

c. Her quality of life should she survive. If the probability was that this would be poor then it would not seem right to subject her to treatments that caused significant pain or suffering.

d. Our own values and beliefs.

Will active treatment prolong the life of our child?

The evidence on whether taking an active treatment approach prolongs life in babies with Trisomy 18 is unclear. Perhaps due to the fact that most infants with confirmed trisomy 18 are managed using a palliative approach, and that there remain ethical questions about offering a more active approach to treatment, the research available is very limited.

A small 2006 Japanese study (Kosho et al) reported on 23 infants with Trisomy 18 who were managed intensively. The median survival rate of 152.5 days and the survival rate at 12 months of 25% were considerably higher than the survival rates reported by Wu et al (2013) in England and Wales where palliative care is standard practice. Median survival in this case was only 14 days, and survival at 12 months just 8%.

Bruns (2010) conducted a small online survey of the parents of 21 infants with Trisomy 18 (eight of whom died prior to the study). All of these infants had been offered neonatal treatment beyond palliative care, although not all had been managed intensively. The survival rates were extremely good, with non-survivors reaching an average age of 37.5 months; survivors had attained an average age of 103.3 months at the time of the study. However it is likely that these high survival rates reflect selection bias.

Janvier et al (2012) interviewed the families of 272 children, around 25% of whom had had full interventions 'as for any child' and 75% of whom had been offered or chosen a 'comfort care' approach. Survival to hospital discharge was similar for both groups but survival at 3 and 12 months was significantly increased in the full treatment group. Of infants in the full treatment group, 68% were still alive at 3 months, 53% at 12 months and 24% at 5 years. The figures for those in the comfort care group were 48% at 3 months, 30% at 12 months and 16% at 5 years. The high overall survival figures reflect selection bias (families were recruited via social networks and support groups) but this is the only study comparing survival outcomes between children with Trisomy 18 and 13 managed intensively and those managed palliatively. The significantly better long-term outcomes for those managed intensively is notable.

Conversely Goc et al (2006) reported on 20 infants with a diagnosis of trisomy 18 admitted to the NICU (eight were diagnosed prenatally). Some treatments were withheld once diagnosis was confirmed, including complete corrective cardiac surgery and aggressive management of cardiac and respiratory failure. 30% of these infants were discharged from the NICU; their median survival time is not mentioned. Infants who died had a median survival time of 20 days, not significantly higher than the median survival in the UK population study (14 days) (Wu et al 2013), although if the survival time of infants who were able to be discharged from the NICU were included, it is likely median survival time would be longer.

Another factor we considered is that postnatal diagnosis is known to be a factor in increasing length of life. This may be because infants with a postnatal diagnosis are less likely to have other complications (eg cardiac defects), or it may be that until Trisomy 18 is confirmed, infants are likely to be managed intensively according to their presenting condition.

Looking at the available evidence regarding whether active/intensive treatment prolongs life in infants with Trisomy 18, in every study, the infants given active treatment survived longer than the median survival times for UK infants with Trisomy 18 (almost universally managed with a 'palliative care only' approach). Studies were generally limited and small, and suffered from selection bias, but there was a clear trend toward prolonged survival in this group, sometimes significantly prolonged. In the only study to compare infants managed using a palliative care approach and infants managed actively, again, infants managed actively had significantly better long term survival rates.

We are convinced that active treatment will offer our baby a better chance of longer term survival: the research of Kosho et al (2006) and Janvier et al (2012) suggests she may have around a 20% greater chance of celebrating her first birthday. Our decision about whether to opt for an active approach therefore depended on whether this approach would cause such significant pain and suffering as to outweigh the benefits of prolonged life.

Will treatment cause 'pain and suffering' and if so where does the balance lie between the 'benefits' and the 'burdens' ?

It is undeniable that intensive neonatal care causes both pain and suffering to the infants subjected to it. To what extent the pain and suffering can be relieved by the use of

pharmacological and non-pharmacological pain relievers and developmental care practices is not entirely clear. The decision to subject infants to intensive care should only be made therefore when the burdens of pain and suffering are outweighed by the benefits of care being provided, in short when neonatal intensive care is in the child's 'best interests'.

In our baby's case, there are conflicting opinions on whether the benefits of possibly prolonging her life are outweighed by the burdens of neonatal intensive care. Our opinion is that, while the research available is limited, it does indicate that her life is likely to be extended by the provision of intensive care, possibly significantly extended.

There is a strong moral and legal presumption in favour of prolonging life even at some cost, and for us, if provision of intensive care will prolong our baby's life, we would like to take that chance, even though we are aware that it may cause her some pain and suffering. We would do our utmost to minimise pain and suffering in every way we could, and would continually assess whether treatment was still appropriate. We would likely discontinue treatment if it was causing severe, unrelenting pain or if it seemed that it was unlikely to prolong her life.

In Janvier et al's (2012) study of 272 families with children with Trisomy 18 and 13, 89% of parents who opted for intensive treatment had no regrets. It seems to us that parents who watched their child endure significant pain and suffering are likely to express regrets. The evidence suggests that families who opted for intensive treatment tended to feel that the burdens were outweighed by the benefits. Their lack of regret is also in contrast to the experience of parents in the same research who opted for palliative care, of whom 31% expressed regrets that they didn't explore more interventions.

Neither we nor the medical team are in a position to assess our unborn baby's wishes. We feel that when the benefit/burden ratio is so uncertain, we as her parents should be able to make the decision after discussion with her medical team. This is the presumption used for extremely premature babies in whom poor outcomes are likely (Nuffield 2007). For example, at 22 and 23 weeks' gestation, resuscitation is only attempted and intensive care offered when parents repeatedly request it after discussion with a senior neonatologist. Outcomes for 22-week gestation babies suggest that only 1% survive to discharge from hospital, and for 23-week gestation babies that only 11% survive this long (Costeloe 2000). In comparison, Goc (2006) indicated that 30% of babies with Trisomy 18 were discharged from the hospital after intensive care.

All babies with Trisomy 18 are likely to have severe disabilities, but this is not dissimilar to the outcomes for extremely premature infants. For 23-weekers, only 7% of survivors had moderate, mild or no disability at age 6 (just 1% had no disability). For 22-weekers, no infants survived without severe disability at age 6. It is unclear why legally and ethically the presumption should be in favour of parental choice in these cases (Nuffield 2007) but not in the case of an infant with Trisomy 18. The pain and suffering caused by neonatal intensive care is unlikely to be different in either group and outcomes are very similar.

If our baby survives, will she have a good quality of life?

Another argument for the withholding of active treatment from babies with trisomy 18 is that even if treatment would prolong life, their life would be of such poor quality due to their significant disabilities that their survival in itself would constitute such a 'burden' as to outweigh any 'benefits' from active treatment. Parents who have received diagnoses of trisomy 18 report being told that if their child survived, she would 'live a life of suffering', 'be a vegetable' or 'ruin their family' (Janvier et al 2012).

Fenton in 2011 in a personal commentary after attending a SOFT (Support Organisation For Trisomy 13/18) conference mentioned that families of children with Trisomy 13 or 18 were averse to hearing talk of quality of life and felt it was often used judgementally by the medical profession, sometimes to deny their children care.
In a study of 272 families with children with trisomy 18 or 13, 97% of parents of surviving children reported that their child was 'a happy child' and 86% of parents of children who had died reported that their overall experience of their child's life was positive (Janvier et al 2012). Smaller studies have also reported that parents of surviving children feel that their child has a good quality of life (Baty et al 1994, Bruns 2010, Kosho et al 2013).

Studies of development also suggest that children with trisomy 18, while profoundly delayed, do acquire a range of skills. Baty et al (1994) noted that all children could recognise their family and smile, and that many children were self-feeding, sleeping independently and using a walking frame, and were able to communicate with signs, crawl, follow simple commands, recognise and interact with others and play independently. Kosho et al (2013) also noted that children with trisomies 18 and 13 interacted with parents and siblings and enjoyed quality family time.

We do not feel that the available evidence suggests that our baby's medium- or long-term quality of life would be impaired sufficiently to justify withholding treatment. It is important to avoid making the assumption that a life with profound disability is one that is not worth living. We have also considered for ourselves whether we could provide our baby with what she would need should she survive, and while we recognise that caring for a child with profound disability is very challenging, we feel we are in a situation in which we are able to do so. We have made contact with other families raising their children with trisomy 18, ranging in age from newborn to 20 years old plus. Their experiences have reassured us regarding quality of life and have also given us an insight into the challenges and difficulties we may have ahead of us if our daughter survives, as have [the mother]'s experiences as a midwife working with babies with disabilities and their families.

Considering our own values and beliefs

Obviously our own values and beliefs have played into our decision-making about the care we would like for our baby. We recognise that there are a number of valid approaches to the care of a baby with trisomy 18. We also recognise that the balance between the 'burdens' of intensive care and the possible 'benefits' of prolonged life is very difficult to determine, and every individual, including ourselves and the medical team caring for our baby, will be influenced by their experiences, reading and values.

We both share a strong presumption in favour of prolonging life. We would ourselves likely accept intensive and painful treatments to prolong our own lives, even if the chances of longer term survival were small.

We also share a belief that disability, however severe, does not render a life less valuable or less worth living. Prior to conception, we had discussed what choices we might make if we had a child with a disability and we felt that, in the event of prenatal diagnosis, termination was not an option for us and that we were both willing to care for a child with a disability. We are also fortunate to be in a position to do so. We do know it would be exceptionally challenging, and even that we perhaps cannot actually yet comprehend quite how challenging. We both believe that treatment should not be denied on the basis of a disability but should be decided based on the clinical condition of the patient.

Our primary consideration is always our child's best interests, and we believe that as her parents we are best placed to determine these. We are open to reconsidering our decisions throughout our journey with our baby Her condition, advice from her medical team and our own perception of how

she is doing are all likely to influence our choices. However, we feel strongly that the final decision should be made by us and not by the medical team.

We also have considered how we would feel if our baby were to die despite active intervention, especially if her death was not as 'good' as it could be. This discussion has also been raised repeatedly by our medical team and we have considered it carefully. Due to our belief that life is valuable, and that our baby's life is valuable, we want to make every effort to prolong her survival while continually weighing the cost for her in terms of pain and suffering. If, after consultation with the medical team, we were to decide that a particular course of treatment was right for her and she then did not survive it – for example if she died soon after birth despite active resuscitation attempts – we feel that we would cope better knowing that we made a well-informed decision, aware of the risks, and that we did what we considered the best for her.

We know we would find it very hard to live with our baby being denied treatment we felt would benefit her, that we were denied the opportunity to do what we felt was right for her. We know that, while generally parents feel they made the right choice for them regardless of whether they chose palliative care or active treatment, parents who choose palliative care only are more likely to have regrets (Janvier 2012).

In Summary

- Limited evidence suggests that an active approach to treatment, including intensive resuscitation and providing clinically needed surgeries, improves survival rates to 12 months (and possibly beyond) in children with trisomy 18.

- There is a strong legal and moral presumption in favour of prolonging life even at some cost in terms of both pain and suffering and resource use.

- Where the benefits of possible life (with or without severe disabilities) and the burdens of neonatal intensive care are finely balanced (for example in the case of extreme prematurity), it has been suggested that parental decisions, after discussion with the medical team, should be deferred to. It is difficult to see why this should not be the case for parents of babies with trisomy 18.

- Quality of life is reported to be good in survivors with trisomy 18 despite severe disabilities.

In short, we feel there is insufficient evidence that restricting treatment to palliative care is the only ethical approach to management of a baby with trisomy 18. Emerging evidence suggests some benefit to an active treatment approach and that survivors almost universally have a good quality of life. When there is such limited and contradictory evidence, it is not acceptable for any hospital trust or individual physician to dictate which approach should be used. This decision should be made by the parents after a full and through discussion with the medical team and should be revaluated by the parents and the medical team throughout the baby's life. Generally parents are the best arbiters of their child's interests.

We appreciate the Hospital Clinical Ethics Committee meeting to consider our daughter's care in such a timely fashion, particularly given the high risk of prematurity associated with trisomy 18 and therefore the urgent need for a care plan to be put in place.

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