

Children who are Long Term Ventilated – Pathfinder Project

Engaging with Families and Children / Young People

*An independent quality improvement review
by the Patient Experience Network*

For
NHS England

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September 2014

Patient Experience Network
Re:thinking the experience



NHS
England



*Engage, empower, and hear patients
and carers at all times.*



**A promise to learn– a commitment
to act:** Improving the Safety of
Patients in England

National Advisory Group on the
Safety of Patients in England

August 2013

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Thank you

We would like to thank the families for agreeing to be interviewed and for giving their time and views and so generously.

Introduction

The number of children and young people in the UK requiring long-term ventilation (LTV) has increased significantly in recent years, from one in 1975 to an estimated 1400 in 2013.

Children who are long-term ventilated have been found to have a significantly lower health-related quality of life. Supporting these children to live as normal a life as possible at home with their families brings many benefits, and any risk involved must be balanced against the benefit to the child of being at home rather than in hospital long term.

Children and young people on long-term ventilation require the most complex care that is given outside a hospital environment and there are significant risks involved in looking after a child on long-term ventilation in the community.

With this in mind NHS England Specialised Commissioning are currently piloting a Pathfinder Working Group model as a way of developing end to end pathways of care for children who are long term ventilated which integrate across commissioning boundaries.

The outputs from the Pathfinder Working Group will include a national set of standards, and recommendations for pathways of care for these patients. As part of this “Toolkit” they want to include a sense of what families experience on their health care journey.

The Patient Experience Network (PEN) were delighted to be selected to conduct a series of 6 interviews with families of children and young people who are long-term ventilated over the

summer months with a view to producing some recommendations and vignettes that will be incorporated into the final report. *(See appendices 1 and 2 for more information)*

Approach

Following a briefing call the PEN team undertook some limited desk research, reviewed information provided by NHS England and WellChild, including the results from two pieces of work recently completed (top three issues, Prime Ministers exercise) and examined the Standard and Pathway. PEN developed an interview guide and then, with interviewees provided by WellChild, completed 7 interviews with family members (including carers) of children on long term ventilation. These interviews were conducted either in person or by phone and ranged from 60 minutes to 270 minutes. They were primarily completed over July, and August with a couple being undertaken in the first week of September due to availability. We also received a document from another parent to review as part of the project.

Despite our intention to ensure that we included the voice of children and young people in this report on this occasion it has been purely parental and carer input. With a longer time scale further work with children and young people specifically could be undertaken.

These families represented a broad range of experiences – across England, from the terminally ill to the soon to be non-ventilated, from children still to get home to those who have been home for years, from babies to young adults, those with extremely rare conditions to those with more familiar conditions and so on. All interviewees were mothers although in many cases father had made notes to be included or had briefed their partner accordingly. *(See appendices 3,4,5,6,7,8 for further information including an anonymised list of who was interviewed)*

Executive Summary

In its current form the standard delivers a range of valuable guidance and covers many of the key issues. From our research and interviews **it is evident that overall there is not enough focus on the emotional journey and needs of all involved – parents, carers and the children.** Parents perhaps have the most evident needs but we should not forget the needs of the others.

In this report we have highlighted the recurring themes that have come out from our conversations. This report is not intended to understate or, worse, undermine the great work that is being done today to care for children with long term ventilation needs, rather it is intended to robustly reflect the views of the parents involved to provide input to further improve the proposed standard and pathway.

Themes:

- Improve the handling of a diagnosis – ensure the professionals have researched it fully and deliver the news appropriately
- Support parents to better understand what to expect from the beginning and provide more information before the tracheostomy – parents need to feel they made an informed decision about this life changing operation and they specifically need to feel that it is one of which they were in control of
- Listen to what parents say – they are the expert on their child. It is ok for staff to say “I don’t know – can you tell me?” They do not feel listened to when it matters most
- Parents should be given the choice to be involved as an expert partner – they can be part of the medical team, but not all parents may want to be – it should not be assumed they will

- Parents have to be strong and push to make things happen – advocating for their child - this can be very difficult when also trying to cope with a very sick child
- Make sure people get the flex in their care package they need, with consistent carers – this is really important for children and young people (and older patients who have young cognitive ages)
- There is a huge inconsistency in the packages being offered – arguably rightly so as there is a huge difference in the support required - but sometimes the financial numbers feel like they are being plucked from the air and the process puts people under undue stress
- Personal budgets are seen as a potential solution to address this – although not all parents are aware of whether they can have a personal budget and feel the process can be very complicated and slow to set them up
- It goes without saying not all children or parents are the same – so there needs to be flexibility in the pathway and approach to reflect this.

More specifically:

- Make sure people can get home quickly without unnecessary delays – removing the barriers e.g. equipment, care package, trained carers etc.
- There is a constant worry about the carers – both for care packages and personal budgets – who is going to be looking after their child? – and are they doing it right?
- Carer staff turnover and the consequent competencies and training needed is also a major concern

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- Some nurses can easily undermine parents' confidence – e.g. get nervous themselves when doing tracheostomy tube change, overly concerned about discharge
 - Team leaders who are taking the decisions e.g. signing off staff as competent are not always themselves competent
 - One single point of contact – parents talk about one person to go to who will manage everything. In reality one could argue that if the system worked they would not need to do this - it is a consequence of a stream of poor experiences
 - 24 hour access/ continuity – e.g. typically the ventilation team were not available on the wards in the evening or the weekend; getting hold of someone who knows in the community
 - Remove the silly hurdles, it is difficult enough without them – e.g. loaning equipment, removing used medical supplies
 - Disabled badges when under 3 – critical for children with ventilation needs (and probably others as well)
 - Need a proper vehicle to transport LTV children – lots of space for wheel chair and extra carer(s)
- they are in control. In the long run this may have a positive impact on overall costs – due to the savings incurred from a reduction in stress related illness etc.
- Remember that events may be 'run of the mill' for clinical staff - they are far from that for parents / carers. Coping with an LTV child is a life-changing experience and families need proper preparation and support
 - Retaining flexibility to respond to the varying needs of these families is very important – there is no one-size-fits-all solution and any pathway of care needs to have flexibility built in
 - Address issues with diagnosis – extended timeframes and inappropriate delivery by consultants
 - Provide one point of contact for parents / carers – someone who will take up the case, co-ordinate the various groups and specialisms, provide a bridge between hospital and home and provide essential support at times of stress
 - Ensure that care packages are properly explained to parents /carers and that they are involved in their preparation from the outset. The feeling of helplessness experienced in the early stages is very debilitating and can be reduced with greater involvement
 - Listen to parents and carers – they know their children better than anyone and working with clinical and nursing staff is more beneficial to both patients and parents
 - Have a can-do attitude rather than putting barriers in place – parents and patients benefit from an element of normalisation e.g. arranging for swimming, family holidays, boat trips and days out in the car

Recommendations

- Parents seemed to find the opportunity to share their experience with an independent group valuable and were keen to have their views included. Further work should be done to ensure we do incorporate the voice of children and young people.
- Reflecting more the emotional needs of the family members is essential to ensure they feel they have been part of the decision – and that

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- Get the 'simple' things right – staff rotas and maintaining continuity of care; ensuring that the correct equipment / consumables are delivered at the right time; help with disposal of used consumables;
- Ensure a level of consistency across the regions and learn from good experiences – much of the report has focussed on negative experiences but there are positive ones which should be identified and rolled out e.g. community liaison in Cardiff
- Training of staff – provide fully trained staff for home care
- Put together a structured training plan for both staff and parents/carers and ensure that care plans, Standard Operating Procedures (SOPs) and preferences / individual idiosyncrasies are 'published' and communicated to all
- Managing transitions is vital - transition from Intensive Care Unit (ICU) to High Dependency Unit (HDU) to home, transition from 'child' to 'adult' at 18, bereavement (looking after a very sick child 24/7 to 'nothing')
- Provide disabled badges for under threes (a parent reported that there needs a change in the law*) and provide for specialist vehicles
- End of life and bereavement care needs careful and sympathetic management - for example - removing equipment within days may not be appropriate, expressing sadness at loss almost certainly is
- It is also worth mentioning here that a few interviewees commented on the generosity of their employers in fund raising and general support (not necessarily something that standard and pathway can do anything about but it takes some of the stress away from coping with such sick children and providing for them)

- Parents need support across four different areas:
 - Professional support
 - Emotional support
 - Practical support
 - Experience support

The pathway and standard needs to reflect all these areas

- The children and young people have three sets of needs:
 - Medical/clinical
 - Social/emotional/independence
 - Educational/ developmental

These all need to be provided and again the standard needs to cater for these

**The law was changed in June 2011.*

https://www.gov.uk/government/uploads/system/uploads/attachment_data/file/212551/blue-badge-scheme-local-authority-guidance-paper.pdf

Children under 3 are eligible for a Blue Badge if they have a specific medical condition which means that they:

- must always be accompanied by bulky medical equipment that cannot be carried around with the child without great difficulty; and/or
- need to be kept near a motor vehicle at all times so that they can, if necessary, be treated in the vehicle or quickly driven to a place where they can be treated.
- Bulky medical equipment includes: ventilators, suction machines, feed pumps, parenteral equipment, syringe drivers, oxygen administration equipment, continual oxygen saturation monitoring equipment, casts and associated medical equipment for the correction of hip dysplasia.

Observations Across Key Phases of the Journey

Below are the key steps in their journey as reported by parents:

1. First hospital Stay and Step Down from ICU to HDU

“When we came out of intensive care we went into no-mans land”

In our cohort, most children were admitted as an emergency onto the ICU – many then moved hospitals due to the limited expertise locally. Often a diagnosis has not been known until later on – sometimes after the ventilation. These are children with a range of diverse and at times very rare conditions.

In ICU parents noted there is 1 nurse per child. Conversely in HDU there is 1 nurse to 2 - 4 children (depending on the hospital). In each case where the parent described this step down they commented that they felt that some of the nurses in HDU seemed rushed/ inexperienced/ over-worked and suddenly they just expected the parent to do the majority of the caring – from simple observations and handovers to other staff, to suction and in one case resuscitation. Parents did not always want this responsibility and the step down from the high level of 1-2-1 care in ICU made this even more marked. Parents felt they were not given the option – nurses just assumed and indeed expected the parent to do this. Several parents commented that through their interventions their child was still alive.

Even if you accept that some of these parents are perhaps understandably over-protective and maybe alarmist, this is a theme that comes through again and again. And whether it is true or not it is certainly their perception of the situation.

Often the ventilation team are not there in the evenings or the weekends and parents report that it is hard to get other nurses interested. They would like hospitals to provide 24 hour access to these vital resources. In some cases while on HDU parents said *“they gave me the feeling we were blocking beds”*.



Comments included:

“We felt it would be safer at home for our child - they just did not have the staffing levels – my child was streaming with secretions but there were no staff to deal with it.”

“The inner tube was blocking so I asked a nurse to help – but they were too busy. I had to go outside and call the trachy nurse myself.”

“On the ICU we were only allowed to be there for 4 hours – so I wrote to the CEO to ask them to extend the hours – which they did for all patients under 25 years.”

“When we came out of intensive care we felt like we went into no-mans land - we were put onto a ward with 4 other children and it seemed that all the nurses were either agency or newly qualified – there were no skilled staff on the ward. I truly believe she would not have made it if I had not been there – the step down is just too significant. The expectation on the ward was that the parents will stay and be the day shift – this was the worst part and the most upsetting time.”



2. Tracheostomy

“I would have liked to have been able to make an informed decision”

The majority of parents stated they did not feel fully informed of the impact of the tracheostomy.

Parents comment that the professionals made the parents feel like a trachy is a routine intervention – without taking the time to explain that this is a life-changing operation. For them it may be a run of the mill small simple operation – for the parents it is massive change to their lives - both their own and their child’s.

In most cases parents were not alerted of the full impact of the trachy for example that two trained carers would be required at any point – and ultimately may have to live in their home, two trained people would be needed to go out in the car, the amount of equipment required, that it will take two people to even go into the garden, the implication for swimming, the bath the shower.

Small things can make a big difference for example for one set of parents when the LTV nurse came and spoke with them *“she explored options with us – inviting a second opinion – we felt like we were given a voice and were involved in the decision – we wanted to be able to look our child in the eye and say we had explored options for them. They also took us to see another child on a nippy ventilator. We could see they were a happy child and felt reassured about what it really means”*.



Comments included:

“I attended a meeting to come home and suddenly the consultant said he needed a trachy - there was no time to decide – he went into surgery the next day – it just happened. Then I went into melt down. I would have liked to have been able to make an informed decision.”

“The consultant told us our child was sick anyway so they were just going to do it. And no-one told us what it would be like so we started to imagine the worst – and that it would be forever and our home would have to be transformed into a hospital. Being treated as an adult would have helped as I had done some research and wanted to review options but the consultant almost laughed. We were given no sense of what having a trachy was really like – and that actually we could have a relatively normal life as we can go out as the kit is battery operated, and it is not necessarily for ever.”

“We were in an intensive care situation and they told us they were going to put one in – and said basically that is ok isn’t it? We didn’t understand the environment or the decision we were making – we were in shock and didn’t have a clue. It didn’t get explained particularly well – we were in intensive care - it was life or death – no one was thinking of the long term view. But nevertheless no one took the time to discuss it with us. In reality this may have been the best possible way – perhaps they do need to be blunt and just do it.”

“For us we had to have the trachy – there was no option –as the Endo-Tracheal tube (ET tube) had been in for weeks but we did not anticipate it would be for the long term. When it was done we could see her face and this was a positive.”

“We always believed the hospital was doing the right thing.”

“We were terrified but have found we can be mobile and it is now part of what we do.”



3. Discharge

*“We were made to feel
unwelcome on the ward”*

When describing the discharge process all parents spoke about this being slower than they hoped. Some parents were keen to get their child home quickly having been told they would not survive beyond their first birthday, others were concerned that their child was not getting the care they wanted on the ward, others were looking to give them a more homely environment, and others were simply keen to get back to a normal family life. Whatever the stimulus all felt the process was flawed. Delays were primarily due to sorting out the care package/ personal budget, recruiting and training the carers and organising the right equipment. During this delay several parents were made to feel unwelcome on the ward – almost as if they were taking up a valuable bed.

Some parents were extremely concerned about leaving the hospital without having a package in place – and that the commissioners may take advantage if it was not sorted and pull the hours – others were unwilling to let a lack of package get in the way of their ambition to get their child home.

Whichever the stance, what was consistent was the challenge and delay in getting an acceptable package in place – with only one family commenting that this was relatively simple.



Comments included:

“Several times I just stood by the bed and wanted to take my child home even though I knew it was unsafe! We would sit there for hours with our backs to the ward looking out of the window –looking out because we could not leave the ward – there were just not enough nurses.”

“I did not know what we would need when we got home – some people suggested 7 nights and 4 days – it felt like a lot but I wanted to understand other people’s perspectives.”

“The last 3 months were most difficult – we had a phased discharge in the first month it was great and then in the next 2 months we had the paramedics and the nurses just sitting in our room and then everyone got very upset when it came time to go back to hospital – we could have looked after her at home as we were ready as a family but the carers were not trained and ready so we were not allowed to take her home.”

“When the day came no-one came to say goodbye - we had been on that ward for months.”



4. Home

*“Home is home – it needs to be safe –
but it must be a home”*

Perhaps surprisingly, although clearly significant changes had to be made to homes to welcome their respective children home, this was a typically relatively low key part of the conversation – with ground floor extensions, ramps, widened doors, extra sockets all being put in place with relatively little comment. Certainly in no case was this cited as a cause for delay in discharge. This seems to conflict with some previous observations and may be worthy of further discussion. The main focus of the conversation was around other key areas as described in this document. Having said that access to grants/funds was not always simple and in some cases ramps had to be adjusted and doors widened after the child had returned home and in another case the wrong bath had been installed.

Finding, training and retaining good quality carers is an ongoing issue for all. Parents are looking for carers who are competent, confident and right for their child. One parent spoke of a carer who came to look after their baby and they had never even changed a nappy before.

They want consistency and continuity in their carers as part of their “family” who know their child and their child knows them and so is at ease with them – but often have a pool or bank of different carers. Many spoke about refusing to allow some carers to look after their child *“we have had 3 members of staff who I have refused to have back – they were falling asleep at night, or just not listening and doing it all wrong for my child”*.

They want carers who both understand the medical needs and also know their child – much as one would when entrusting a child to a nursery nurse – you want to be sure they know what your child likes to do, how they communicate, when they are happy/ frustrated etc. One commented *“he has his preferences and I think he is quite a good judge of character too!”*



Comments included:

“When you have a consistent team of carers they get to know what your child likes, how you like to do things and it becomes fairly easy.”

“They want to medicalise your home – the carers were told they had to wear uniform – but my child did not want them to – she became distressed – so they would bring their own clothes and change into them when they went out – but they had to change back into the uniform when in my home.”

“The changes since we have got home are amazing – which shows us that we made the right decision and the timescales in the hospital are far too long.”

“Within weeks of being home she was perkier and happier.”



5. Carers in the home

“I felt guilty because I was not doing things for my son”

As stated in the previous section, all parents talk about the critical importance on being able to choose who is the carer for their child – and many observe they had to refuse entry to some – who they did not feel confident or comfortable with. When it works the carers become an integral part of the family – and are trusted – even sometimes being relied upon to over-rule the professionals.

However, having people in the house all the time raises many concerns – and parents are not sure of the appropriate etiquette.



Comments included:

“I felt weird about the etiquette – and asked the group on Face book. I found it difficult to let go and felt that I needed to be with him all the time – but really it is like having any child- sometimes you need to go and do the ironing – but at times it felt really difficult because I was making them the child minder too – and I felt this was not OK.”

“Having people in the house was a concern at the beginning but now I am very comfortable. We have the flex to ask them to swop a day to an evening. Everyone would benefit from having a similar set up – a dedicated team – allowing you to behave like a family and have some time out.”

“With the personal budget (you can only have this if you have been in the system for 3 months) – but with this it is fine for the carers to be on their own with my child – others are not allowed to do this. We have quite a lot of freedom and flexibility in our care package. This is really important for us – and it should be available for others too.”

“Some carers are amazing – they know what she needs and can be great playmates. They are limited by their role – they could do as much as the nurses – we are piloting trying to get carers to do the nursing role.”



6. Re-admission/ local medical support.

“The local hospitals do not know how to deal with a trachy and LTV”

As parents of very sick children – being “blue-lighted” back to hospital is a very real possibility and as part of the package the alternatives have to be discussed and fully explored.

Parents talk here about their child being refused entry, packages being removed, departments being unfamiliar with the kit, the condition, with children. At a time of incredible heightened stress knowing you are not welcome and your options are restricted must be a massive pressure to deal with.



Comments included:

The GP refused to see her – they were very open and said everything she has is too complicated – it was honest but not very helpful.”

“The local hospitals do not know how to deal with a trachy and LTV. If he has to go in they will not allow the carer in - saying they are paying twice for the carer and the nurse so when they are back in hospital the package is removed. So now they will deal within A&E until he is stabilised to come back home.”

“The local district hospitals were not equipped to deal with us.”

“When we were in an emergency setting they had not seen a nippy ventilator before – they are not familiar with the paediatric set up.”



7. Transition

“Although 18 she is still a child”

The move from children’s to adult services is a very real concern for all parents of children with a chronic illness. This is exacerbated by the complexity of the child who is long term ventilated – and indeed for those where they are also cognitively impaired.

One child turned 18 while in a specialist hospital. For this patient although she was 18, her cognitive age was 10/11 and she was very small for her age. It became apparent that the local children’s hospital did not want the young person back and this caused a wide range of challenges for the parent – from expecting to be allowed to stay in the cubicle with their child as they had been used to on the children’s ward, to expecting to be a part of the support team, to being allowed to stay when she collapsed.

An area that perhaps the guidance can provide support is highlighting the need to set up power of attorney fairly early on. It took one family 10 months to do this due to the complexity of the health issues and without it their young adult would have been even more vulnerable

One parent of a 5 year old is already concerned about the prospect of transition



8. End of Life

“It makes you focus on the dying not the living”

This is an incredibly difficult area to get right – and there can be no one simple set of guidelines. What is clear though is that is in an issue that deeply concerns parents.

In several cases the children are terminally ill – in all these cases the parents were very focussed on making sure their child had the best quality of life, focussing on living.

Parents expressed deep concern about the future for their child – there is a feeling that many professionals do not agree with the long term ventilation approach and feel that these children and young people should have the ventilation removed.



Comments included:

“She was transferred to an adult ward – my husband and I took it in turn to stay with her – and they were not used to this on the ward and it took time for them to accept this – but although 18 she is still a child.”

“They need to understand that parents are integral to what is going on - if she collapses they need to be able to stay - it is important for you as a family – she needed people she knew there – but in adult settings they did not see that.”



Comments included:

“There is often too much focus on end of life care – they kept talking about preparing and thinking about withdrawing medicines – when you talk about end of life care it makes you focus on the dying not the living.”

“We are worried for the future of our child and for other children in similar situations.”



9. Bereavement

“This was too soon for us!”

This is an area that is not currently covered in the pathway but sadly will be an important stage in the journey for many of the families of LTV children. One family had been through this process and they observed that on the second day after their child’s death the agency came to collect the equipment.



Comments included:

“This was too soon for us – they took everything away even the cut trachy tapes – I wish I had kept those.”

“The carers were mortified by the way we were all treated - the nurse told them to cheer up and get on with it. One had known our child for over 3 years! The carers took it upon themselves to help us arrange the funeral.”

“There was nothing from the care team manager – no letter, nothing.”

“You have to remember our house had changed from being Paddy’s market to nothing over night – we used to have 2 carers, my child, my husband, 24/7 and the equipment, the noise – I used to go to sleep to it, hospital appointments letters, forms then nothing.”



Other Issues That Are Raised

Across the various experience there some themes that crossed several steps in the journey

1. The emotional roller-coaster

“At times I feel there is no-one to help”

All parents spoke about the ups and downs of their experience. Some have felt suicidal at times, and at least two parents have been suffering severe depression. One father was made redundant – understandably according to the family - due to the amount of time he needed off, and one went bankrupt for similar reasons. The stress placed on these families is unimaginable. This is a truly challenging set of circumstances for any parent to have a sick child at home with long term ventilation needs, however, so many additional hurdles are encountered – ones that frankly they could do without. Many try to remain optimistic and focus on the positives but all have to battle for the best for their child and at times struggle to cope.



Comments included:

“At times I feel there is no-one to help”



2. Focus on the possible

“A good day is when my child ticks off another first”

Parents on the whole were very keen to share their experiences and generally had a very positive outlook on life – preferring to look at what can be done and setting challenges to improve their child’s quality of life.

One parent was arranging for their child to go on a boat trip – and this child already goes swimming every week despite having been told this would not be possible.



Comments included:

“A good day is when my child ticks off another firstor another first is arranged e.g. we have started to go swimming on Wednesdays –finally the whole family got to go with my child in an inflatable ring. He loves the water and the freedom of movement.”

“We have a battle with the care team - she is fully ventilated and peg fed and they want her to stay in – she is a child and I want her to go out every day and have a normal family life – with her sister. We now go out every day – it needs careful planning but we now have it all set up and ready. When she was 3 we got access to a van – we were not eligible for mobility allowance until she was 3.”

“You can live a full life on a trachy.”

“There are not many active fully ventilated children – we have a special tray on her tricycle – and away she goes – it gives her independence.”

“The consultant was brilliant – they focussed on quality of life and the living.”



3. Role of the expert parent

*“I just want people to listen
and learn from me”*

As parents they have become expert carers for their child – whether they wanted to or not. They feel they should be given the choice to be involved and where they wish to be they should be considered an expert partner and a key member of the medical team – they know more about their child and their needs than anyone else.

What can be particularly frustrating for parents is when people are following text book or standards rather than understanding the actual patient’s needs – and indeed listening to the parent (or carer) who really does know best e.g. “it is OK for his SATS to be at 92 for some timedon’t panic and interfere all the time....”

One parent observed how difficult it can be to bond with a child that may die and that you have had to resuscitate – for them the burden of having done CPR on their own child is immense and one they do not wish to repeat – yet because of their background they are often expected to do CPR. They ask is it ethical to demand that a parent is responsible for the resuscitation of their own child?

Working parents feel guilty about wanting a career. And the system does not feel like it is set up to support them – “the care team are not allowed to support us to work. School does do after school clubs but there are no funds for a nurse to cover this”.

And it is not just the system it seems that some parents also do not understand why some people would want to work rather than care for their child and at times have been very vocal about this view with at times reportedly viscous comments on facebook. Some parents are frightened to voice their desire to work – but why shouldn’t they? Parents want to be good role models for their other children and for some working is an important part of this – they feel there are multiple financial and political barriers to being able to achieve this.



Comments included:

“I have learned virtually everything there is to know about my child.....I just want people to listen and learn from me – not the text books/ sticking rigidly to the text book procedure.”

“I have to be the advocate for my child.”

“The nursing was fairly spartan – and so we had to do the suction ourselves.”

“We have always been very active in the care plans – and are very directive in the care that we want – challenging protocols , requesting equipment etc. – the care team have responded well.”

“We went down the route of the personal budget and it is the best thing we have done.”



4. See the child not the problem

“Our child is a feisty madam! She demands to be treated like a normal 6 year old”

Too often parents feel that the professionals are just dealing with the medical facets of their child – and not seeing the child behind the condition. They are looking for people who see the child first.

This challenge extends to schools – local schools refusing the child due to their needs e.g. extra ramps or schools not providing transport.



Comments included:

“Our child is a feisty madam! She demands to be treated like a normal 6 year old and do the same things as her sister – it is our job to find a way!”

“It is important when selecting the carers that they see the child not just the patient.”

“I realised that in the daytime ideally the carers will also be trained nursery nurses who are trained for trachy support – as young children they are developing so fast they also need to be stimulated by someone who is trained.”

“The staff on the care team are employed to look after her medical needs – not her social needs – we live on a big hill and they refuse to push her up the hill.”

“No one wanted her anywhere – and then for the first time they did not see her as a problem – she was not too small, too disabled, too old etc. – they just dealt with her issues and sorted it out for her – they just did it.”



5. Parents' Perception of the Professionals

“The person who wants to do it makes the difference”

For several parents the diagnosis process and the way the news was delivered leaves much room for improvement. One parent commented “it should never have to happen to other parents”.

In one case the consultant was very “doom and gloom” and the parents felt she clearly had no real understanding of the condition and had made no real effort to find out more before speaking to them. She did direct them to a website for the condition, from which the parent subsequently made contacts and progress in understanding the condition and the prognosis. Having taken time to educate themselves they feel that the consultant did not look deeply enough into the case and had not bothered to understand the possible outcomes,... and were advised to ‘let him go’ as their child would have no quality of life. From their research these parents discovered that there were some 18 year old boys who had never been trachy-ventilated. They also found out children were going to school in their own wheelchairs, and there were examples of children who were ventilated having real fun. Their question was – why can’t my child do that?

One parent recalls that when their child lost their swallow their consultant told them in the corridor “well this is the beginning of the end”.

Parents feel they have to be advocates for their children – but sometimes when they do this they are given the feeling they are being difficult and have an attitude.

Children who are Long Term Ventilated – Pathfinder Project Engaging with Families and Children / Young People

At times conversations can be extremely difficult and on occasion combative as both commissioners and providers refuse to provide for their child made even more challenging as there is in-fighting too across local boundaries and between experts – all very visible and upsetting for the parent to see.



Comments included:

“Oh are you still here – why aren’t you home by now?”

“Some members of staff were amazing – they would come over at the beginning of their shift and ask how you are doing, tell you their name and say if you need anything just let me know - others were just rude.”

“On one occasion they were having a conversation at the end of the bed saying they had colonised his pseudomonas! I was sitting there and they were not involving me in the conversation – so I had to get up and listen – which they did not like – but they got used to it.”

“What we went through is not normal but the staff at the hospital made us feel as if it is and they were very dismissive – and made us feel as if we should just be able to cope. They had the capacity to make the difference.”

“When the day came no-one came to say goodbye - we had been on that ward for months.”

“We have one consultant who is brilliant – he responds any time – 7 days a week. For the first time she was no longer a problem.”

“At times I feel they are threatening me with my child’s life.”



6. It’s the managers that are the problem!

“It is all generated by the manager”

This came through in several interviews – when working well, the carers can be and are amazing – knowing intuitively what is right for the child in their care – being trusted by the parents to look after their child. There problems tend to arise when managers intervene - from simply ensuring the rotas/ shifts are correct to provide the right level of care, to ensuring there is cover and planning for replacements regarding training etc.

“Managers” tend to insist on text book care rather than understanding the patients actual needs. Team leaders who are taking the decisions e.g. signing off staff as competent are not themselves competent. Managers/ medics/ therapists – want to control what is happening- without always having the full information or understanding the situation.



Comments included:

“We have had a year of hell with our care company – and it is all generated by the manager - we were not allowed to be involved in the recruitment of our care staff – being competent is just not good enough for our child.”



7. The essential role of social media/ personal networks

“The on-line group was a lifesaver really for the non-medical stuff”

There is a lack of good information at the time of the need and parents turn to the internet for support – joining relevant support groups e.g. Tracheostomy UK on Facebook, syndromes without a name, post pals, parents of kids with epilepsy, parents of babies with diaphragmatic hernias. These groups are useful to ask simple questions – often quite basic. The value of the groups are they are linked directly to parents who have been there and done it already – and intuitively understand the issues. There are tensions though that manifest and put some parents at unease for example the tension between those who wish to continue to pursue a career vs those who refer to stay at home.

Local parents who have gone through a similar experience are also mentioned as invaluable support – parents met at antenatal classes, on the ward or unit, through the network.

Existing friends often take time to become comfortable with the situation and therefore provide limited support in the short term – finding excuses or reasons not to be there. “At the beginning our friends did not come over – it took them a couple of years to get ‘normal’”. “After a while friends stopped coming because it was a long way to the hospital and parking was too difficult”

Other people who have been helpful particularly include specialist contacts e.g. A&E doctor, chaplain on the ward, the Armed Forces; “I asked the difficult questions of a friend of mine who works in A&E”, “it was the chaplain who saw I was not alright and got help”.

Of course family members particularly grandparents and siblings also are extensively involved in the care of the LTV child. “as a family we have great family and friends”



Comments included:

“I was looking for guidance on trachy dressings and they explained the tapes to hold the tube.”

“One brilliant suggestion from a parent on the trachy group was to have head torches in case of power cuts.”

“This group really helped when my child first came home.”

“The on-line group was a lifesaver really for the non-medical stuff. Some people post all the time. I joined the group a few days before the operation and asked for tips for how to get through this period. My biggest thing was how to get my son home – to sort out the care package etc. What struck me from the answers was how different it is across the areas so no one had the answer for our area.”

“I did not think I would use this group much but I have found it very useful – I read a lot of the posts as often people are asking what I was thinking about! Some people on the group seem to panic too much though.”

“I use the trachy group for advice – silly things like bags for the back of the wheel chair. I spent hours looking and then asked on facebook and was given some really helpful ideas.

Also I was planning to take 2 suction units to school but someone said I should check if the school had one already – which they did!”



8. The elephant in the room

“It was clear that their commissioner did not want to have to pay for the long term ventilation of our child”

Much of the tension perhaps stems from the ever present financial pressures that commissioners and providers are under – a child with long term ventilation needs comes with a significant price tag. What has become evident through our conversations is that this has become a significant omni-presence that is not being addressed. Being made to feel that a bed is being “wasted” on their child, or that their child should be allowed to die cannot be tolerated. Parents will continue to be placed under extreme and unfair pressure – either we do look after child with LTV needs in their homes – or we do not.



Comments included:

“It was clear that their commissioner did not want to have to pay for the long term ventilation of our child and the associated needs, with the CCG suggesting that we, the parents could take on one of the full time carer roles – they have withdrawn the payments.”

“A lot of doctors talk about extinguishing life.”

“I felt under pressure to just give in.”



9. Disabled Badges

“To travel with your child is a vital part of life”

Mobility is a major issue for families – particularly in the light of the fact that children who are ventilated are going home increasingly early – at 3 and 6 months. Although children under 3 are eligible for a badge, parents still comment about being refused a badge and the challenges this

brings. It is vital to quality of life – for parents and child that they have access to the right vehicle and the right access via a disabled badge.

Two families observed that children under 3 are not eligible for a disabled badge.

See page 6 for further information about blue badges.



Comments included:

“Being able to travel around makes huge difference – not being constantly tied to the house, and one room of the house.”

“I believe passionately that to travel with your child is a vital part of life.”



10. Transport

“Vehicles can need extensive modifications”

A couple of families mentioned that a normal family car cannot cope with the needs of a LTV child – who needs 2 people to travel with them, plus space for equipment. One problem is the electrics needed to power the equipment – the voltage is too high to run the humidifier in the car etc. So vehicles can need extensive modifications. One parent wondered why a mobile unit had not been developed.



Comments included:

“We had to wait until our child was three to be eligible for a mobility allowance. In the meantime my husband has had to have a hip operation – as a result of lifting our child and the accompanying equipment in and out of the car for the intervening years.”



11. Remove the silly barriers

“It was bonkers really”

Silly things happen – if something is done/ learned by one family parents question why the system is not thinking about transferability to others in a similar situation.



Comments included:

“I was pushy and took control of discharge – I was on the phone all the time talking to the commissioner, provider LTV nurse etc. The biggest barrier was equipment – I was told that usually it is the carers but I was pushy! When it was time for the home visits the equipment was not available so I asked the hospital to loan the equipment – they said no! I pushed upwards and gained agreement to loan it – it was bonkers really as it was the same equipment as he was using in the hospital – it took another 2 months to get the equipment delivered at home – so if I had accepted the no he would have been in hospital for another 2 months.”

“Extend visiting hours on ICU for children and young people – reflect their cognitive age – not their physical age.”



Project Background

NHS England Specialised Commissioning are currently piloting a Pathfinder Working Group model as a way of developing end to end pathways of care which integrate across commissioning boundaries. Many specialised services, including pathways for Children/Young People on Long Term Ventilation, span several parts of the health system including primary, secondary, tertiary, local authority and education. NHS England wishes to work together with Clinical Commissioning Groups (CCGs) to develop a process for ensuring the effective commissioning of care pathways for the services (specialised and non-specialised relationship) that form the whole provision of care. The Specialised Commissioning, Women's and Children's Programme of Care and the NHS England Commissioning Assembly (Shared Commissioning Working Group) are supporting the Pathfinder for Children's Long Term Ventilation. A Project Group has been established to take forward this work which includes a CCG Commissioning Sponsor, nominated through the Commissioning Assembly, CCG Commissioners, GP's, Tertiary Providers, Local Authority, Third Sector Providers and importantly Parents.

Engaging with Users of the Service

The outputs from the Pathfinder Working Group will include a national set of standards, and recommendations for pathways of care for these patients. As part of this "Toolkit" we want to include a sense of what families, including the parents and the child/young person, experience on their health care journey.

In order to get the essence of this, recognising that families will have very varied experiences, we will be conducting six family interviews over the summer period. We will aim to approach three parents/carers and three children (age 12 and over), who will be offered the opportunity to tell their story. In order to get a geographical spread we will be approaching families across from London, the Midlands and the North of England.

We have successfully engaged with colleagues from the Patient Experience Network, who have a firm evidence base and wealth of experience of talking with families in this way and they will conduct the interviews on our behalf. We are also working closely with the Director of WellChild who will be helping to identify suitable families to engage. The aim will be to approach families from both invasive and non-invasive pathways.

Time-Scales

The interviews will be conducted over the summer months and the Patient Experience Network will produce a series of vignette's which will be incorporated into the final report. This will be completed by early September in order to meet the Pathfinder deadline of mid-end September.

LTV Background

The number of children and young people in the UK requiring long-term ventilation has increased significantly in recent years, from one in 1975 to 24 in 1988, 141 in 1998, 933 in 2010¹ and estimated to be c.1400 in 2013. Noyes (2006, p.8) highlighted the human costs of long-term ventilation and found significantly lower health-related quality of life in children who are long-term ventilated. Parents also identified consequences to long-term hospitalisation of their children including reduced communication, concentration and confidence. Signs of institutionalisation in children's behaviour have also been identified by parents, including shyness and a lack of a sense of belonging (Noyes, 1999, p.446). Linahl and Lindblad (2011) highlighted that limited available time affected the inclusion of families in their work and community with lack of formal care packages being a major contributory factor. Children and young people's thoughts around health and the restrictions of long-term ventilation can identify the extreme frustration felt by some children, for example 'it makes me feel like some kind of dog, chained out in the front yard' (Linahl, Lindblad 2011, p.252).

Children and young people on long-term ventilation require the most complex care that is given outside a hospital environment and there are significant risks involved in looking after a child on long-term ventilation in the community. Supporting these children to live as normal a life as possible at home with their families brings many benefits and any risk involved must be balanced against the benefit to the child of being at home rather than in hospital long term. All risks must be fully assessed, understood by the family and managed appropriately. A balanced approach to risk management is needed if children and young people are to take advantage of the quality of life, educational, social and developmental opportunities of living in the community. Their care must be of the highest possible quality "in a child-centred environment, by staff trained in caring for children" (*Learning from Bristol. The Report of the Public Inquiry into children's heart surgery at the Bristol Royal Infirmary 1984-1995*, Ian Kennedy, July 2001, p2). "Lack of training in treating children and young people may lead staff to treat them inappropriately, however unintentionally. This includes not recognising children's different clinical needs, or not engaging with them in an age-appropriate way or, for younger children, not engaging with their parent or carer appropriately." (*Getting it Right for Children and Young People – A Review*. Professor Ian Kennedy, 2010).

¹ 1975 and 1988: Robinson (1990), 1998: Jardine et al (1999) - All cited in Noyes (1999) p. 441. 2010: Wallis et al (2011)

Introduction to LTV Standards

These Quality Standards aim to improve the quality of services for children and young people needing long-term ventilation. The Standards help to answer the question: “For each service, how will I know that national guidance and evidence of best practice have been implemented?” The Quality Standards are consistent with NHS England’s Service Specification E07/S/c ‘Paediatric Long-Term Ventilation’ and are suitable for self-assessment, monitoring by commissioners and use in peer review visits. They describe what services should be aiming to provide and all services should be working towards meeting all applicable Quality Standards within the next two years.

The Standards have been developed by a Sub-Group of the Midlands Children’s Long-Term Ventilation Network with input from Dr Andrew Selby, Consultant in Paediatric Intensive Care and Long-Term Ventilation, Alder Hey Children’s NHS Foundation Trust and Elspeth Jardine, Ventilation Service Coordinator, NHS Greater Glasgow and Clyde. The Quality Standards are based on and support implementation of national strategies and guidance, including NICE guidance and Quality Standards..

We hope that through the Quality Standards and, at some future date, a peer review programme, the quality of services providing long-term ventilation for children and young people will improve and in particular:

- a. Children and young people who need long-term ventilation, and their families, will know more about the services they can expect.
- b. Commissioners will be supported in assessing and meeting the needs of their population, improving health and reducing health inequalities, and will have better service specifications.
- c. Service providers and commissioners will work together to improve service quality.
- d. Service providers and commissioners will have external assurance of the quality of local services.
- e. Reviewers will learn from taking part in review visits.
- f. Good practice will be shared.
- g. Service providers and commissioners will have better information to give to the Care Quality Commission and Monitor.

Scope of the Quality Standards

Long-term ventilation is defined as "any child when medically stable requiring a mechanical aid for breathing after an acknowledged failure to wean [off the ventilator], or slow wean, three months after institution of ventilation" (Jardine et al, 1998). All ventilation was once exclusively undertaken in children's intensive care units. The vast majority of children who receive ventilation receive it in acute medical situations and, when this situation is resolved, the need for ventilation usually ceases. For a small group of children, however, the need for ventilation remains when the acute stage has passed. For this group of children, usually referred to as 'long-term ventilated children', the last two decades have seen a move from life in an intensive care unit to life at home. Within this group there are different levels of need, as defined in the National Framework for Children and Young People's Continuing Care (2010):

High (Level 1): Is able to breathe unaided during the day but needs to go onto a ventilator for supportive ventilation. The ventilation can be discontinued for up to 24 hours without clinical harm.

Severe (Level 2): Requires ventilation at night for very poor respiratory function; has respiratory drive and would survive accidental disconnection, but would be unwell and may require hospital support.

Priority (Level 3): This includes those with no respiratory drive at all who are dependent on ventilation at all times, including those with no respiratory drive when asleep or unconscious who require ventilation and one-to-one support while asleep, as disconnection would be fatal.

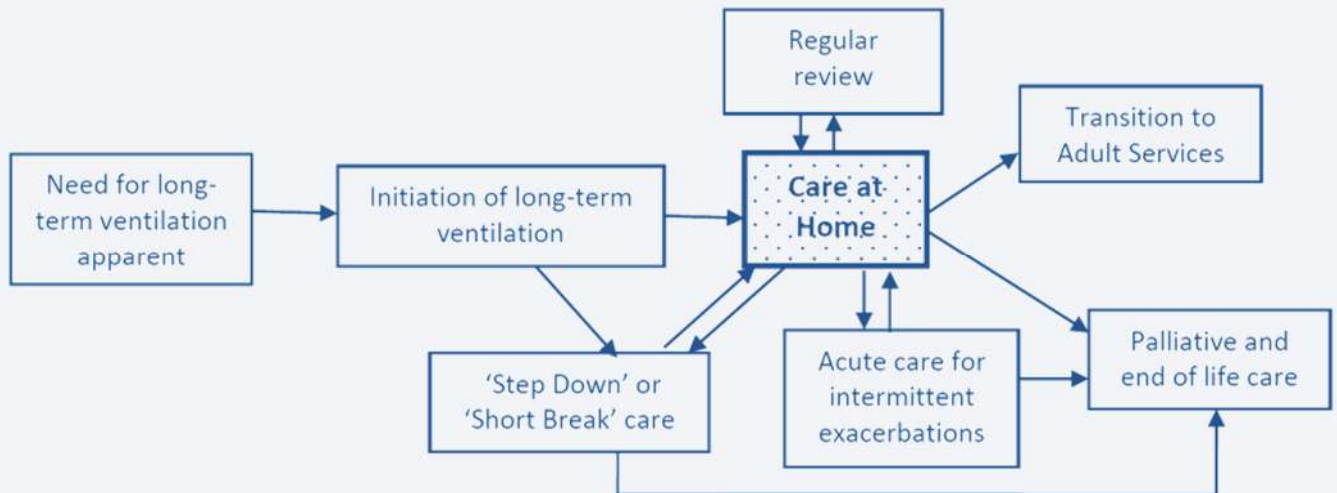
These Standards apply to the care of children with an assessed and agreed continuing care package and those needing level 1 care who do not have a care package. They apply to both long-term invasive and non-invasive ventilation. A small number of Standards, or sections of Standards, apply only to children needing Level 2 or Level 3 care, or to children needing Level 1 ventilatory care but whose other conditions result in a higher complexity of need.

The Standards are about the care of children and young people. Transition to adult services normally takes place between the ages of 16 and 18. In exceptional circumstances transition may be delayed but should be completed by age 21. Flexibility in the age of transition may be necessary while commissioners and providers ensure appropriate arrangements are in place. Some services provide care for adults as well as children and so transition will involve a change of funding arrangements at age 18 but may not require a change of care provider.

Introduction to LTV Pathway

The pathway of care for children and young people needing long-term ventilation is summarised in Figure 1.

Figure 1 Pathway of Care



Throughout this pathway, each child and young person needing long-term ventilation, and their families, should have the information, support and care that they need, in particular:

- A lead consultant from the Tertiary Long-Term Ventilation Service
- A nominated Community Children’s Nurse who will liaise with local services as required and may also be the ‘key worker’
- An agreed ‘Personal Care Plan’
- A review of their care at least annually

Children needing Level 2 or 3 care, and some needing Level 1 ventilatory care who have other medical conditions, should also have:

- A nominated ‘key worker’
- 24/7 access to children’s nursing support who will be able to access medical advice if required

A range of services is needed to support the care of children and young people on long-term ventilation, including transport, education, housing, social care, ambulance and other services. These services are in addition to the wide range of local voluntary organisations, for example, Beavers and Brownies, who help to support social integration. A few children will be unable to return home and will become ‘looked after children’ needing long-term foster or residential care.

Examples of vignettes

The following case studies illustrate different levels of ventilation. Names have been changed to preserve the anonymity of those involved but all have given their consent for the following information to be used in this toolkit.

Case study 1 – Mikael: level 1 ventilation

Mikael was born with Duchene Muscular Dystrophy. At the age of 14 he began to suffer with headaches on waking and generalised fatigue throughout the day. During this period he also developed acute pneumonia and was admitted to his local paediatric intensive care unit where he was ventilated for several days. Following his recovery from this illness and during his subsequent care on a children's respiratory ward, investigations found that he would benefit from overnight non-invasive BIPAP ventilation via a face mask. He was discharged home with a Nippy 3+ ventilator.

Mikael can breathe easily without his ventilator during the day and initially struggled to accept that it was of any benefit to him. He dislikes wearing the face mask and for several months he would only use the ventilator for short periods at night if coerced by his mum.

Over time, however he has come to realise that his quality of life is much better on the days after he has used the ventilator. He has gradually started to use it for several hours every night. Mikael does not require additional support when using the ventilator and does not have an overnight care package.

Case study 2 - Maria: level 2 ventilation

Maria is 17. She was born with severe cerebral palsy. Her condition has deteriorated over recent years. Because of severe scoliosis she now requires level 2 intermittent positive pressure ventilation (IPPV) using a Nippy 3+ ventilator via nasal prongs, for up to 24 hours a day. She is able to breathe without her ventilator but would become very unwell and require hospital admission if she were without it for any significant length of time. She uses the ventilator most of the time; she is only without it for short periods during the day to reduce the risk of tissue damage around the nasal prongs.

She lives at home with her mum and visits her local children's hospice for day care every week. Maria does not have any additional care at home overnight. She is often anxious about her condition and has initiated several conversations with hospice staff, talking openly about her hopes and fears. She enjoys visiting the hospice and also feels that her mum benefits from the break.

Case study 3 – Ameera: level 3

Ameera is six years old. At the age of two she developed acquired central hypoventilation syndrome caused by brain stem lesions of unknown origin. A further complication of her condition led to her requiring a tracheostomy to support her breathing. Following an acute exacerbation of her condition she is now unable to breathe without the support of her ventilator when she is tired, asleep or unwell. She is fully ventilated overnight via a LTV 950 ventilator. Her difficulties are compounded by complex seizures that regularly lead to respiratory arrest leading to a need for resuscitation via bag-valve to her tracheostomy.

Ameera has an extensive care package to support her needs. At home, she has overnight support every night from her local Community Children’s Nursing Service plus care either at school or at home for five days per week. She also has care for two evenings per week, and every six weeks she has two full days of respite. When she stays at the children’s hospice she is cared for by nurses who have been trained to manage her ventilator. There she receives one-to-one support both day and night. Her parents value the break this provides as when Ameera is at home it is difficult for them to have quality time together: They have very little opportunity to spend time alone, especially at night, as she needs to have nurses or health care assistants with her at all times, or one of her parents has to stay with her to monitor her ventilation.

Discussion Guide for Families of Children with Long Term Ventilation Needs

Introduction:

- Thank for taking part in our project
- Introduce self (be ready to share - mum, boys, hopes, fears), PEN and how we are working with NHS England and WellChild
- Explain what we plan to do – have a conversation that we will then use to help us shape the Standards and Pathway currently in development to reflect the carers and children’s and young people’s perspective and experience.
- Explain we also hope to include vignette’s or stories to support the Standard and bring it to life
- Clarify how long we have for the conversation
- Check ok to continue

Background:

- *Discuss* child – focus on the child first – what do they like doing, hate doing, hobbies etc.
- Then ask about the condition - the background and history – as much or as little detail as they are happy to share – e.g. how long has their child been ventilated, why did this start
- How did they first hear about the news
- *Ask* them to describe how this has changed their world

Their journey:

- *Explore*- through open questions and conversation - their journey – from when they first went onto ventilation to now
- *Ask* them to describe a really good day – explore in detail what makes it good
- *Ask* them to describe a really bad day – understand what would make it better

In hospital:

- *Discuss* the hospital stay in detail:
 - When was it, which hospital, how far away, how long were they there etc.
 - How did they feel, what was it like, did they feel welcome and looked after

If they were to have their time again what would they like to be done differently. What would make their time in hospital even better?

During Discharge

- *Discuss* the moment they were given the opportunity to bring their child home:
 - How did this happen, when was it, how did they feel, what steps did they go through, how long did it take, who was involved?
- *Explore*
 - communication across the various teams (what went well, what could be improved)
 - care packages
 - training for them
 - information provided
 - support provided
- What could be done better? What advice would they give another parent who was going through the same process?

First weeks at home:

- *Discuss* the first weeks at home:
 - How did it feel to be the carer?
- *Explore* the following areas:
 - the environment at home (space, sockets etc.)
 - the equipment (training, cleaning/ maintaining, back up kit etc.)
 - communication (local authority, community, hospital, paediatrician, key worker, schools etc.)
 - emotional support
- What went well? What did not go so well?
- If they were preparing a guide what tips would they include for other parents?

Long term ventilation at home:

- *Discuss* (if relevant) the period following the initial few weeks:
 - What got easier?
 - What is still proving very difficult?
 - How often do they see their key personnel
- If they could change on thing what would it be?
- If they could describe one positive about their experience overall what would it be and why?
- What else would they like to discuss at this time that may make a difference to the Standard and Pathway being developed?
- Finally ask if there is anything else they would like to discuss
- Describe what will happen next
- Thank them for their time.
- Close

Specific information to collect:

Parents name, email and tel no

Level 1, 2 or 3

Invasive or non invasive

Name, Sex and age of child

Where they live

Condition

How long they have been ventilated

Are they willing to be contacted again if necessary

Would they be happy for us to write up a vignette to capture their story

Table of Participants

Age	Location	Ventilation	Contact
10	North	Level 2/3 Tracheostomy - ventilated night and most of day	Visit
3	South	Level 3 Tracheostomy - ventilated	Visit
6	East	Level 3	Telephone
23 (<i>cognitive age 10/11</i>)	West	Level 2/3 Tracheostomy - ventilated night and most of day	Telephone
1	Midlands	Level 2 Weaning off ventilator - 4 hours a day	Telephone
2	Central	Level 2/3	Telephone
5	Midlands	Level 3 Tracheostomy - ventilated	Telephone

We would like to thank all parents and carers for giving their time and views so generously

Vignette 1

Introducing Liam

Liam is 10 years old. He enjoys watching TV and together he and his team take the time to watch a film each afternoon. He loves people talking and interacting with him, especially when they dance around and he has a carer, Natalie who is a great dancer and really makes him laugh. The Hobbit and The Lion, the Witch and the Wardrobe are amongst his favourite books. Liam has been recognised for his bravery when he received an Award from Prince Harry. When it is available Liam likes to use the Eye Gaze.

Introducing His Family

Liam's mum used to be a PA and was an active member of her local gym. She was in control of her life. Since Liam became ill she worked for the first six months but since he became very sick she has given up her job to look after Liam. She now rarely leaves the house.

Liam also has a brother – who is at University and visits frequently. Liam's father no longer lives with them. And his mother's family do not really get involved – they have not seen Liam for several years. At times his mum feels very isolated.

Liam does have a team of great carers and his mum has a friend who lives just around the corner whose child has also had a trachy. She has found groups on the web really helpful at different stages of Liam's illness.

Introducing Their Experience

- Liam was a regular child until he was three and then one day he woke up and couldn't walk. At first they thought it was lazy hip syndrome, but over the years he has progressively lost the ability to move and is now peg fed, fits regularly, and has been locked into a mental ability of three years old. He has been in and out of ICU and has had a range of procedures including a tracheostomy. He is ventilated at night and most of the day.
- He was intubated because they could not stop his fitting, and his mum comments "I would have liked to make an informed decision".

Understanding the condition

Finally after many years of not knowing Liam has been diagnosed with Batten disease – a very rare degenerative condition which is terminal. At last his mum knows what the future holds and no longer feels she is in some way responsible for his illness.

Batten Disease is also known as Spielmeyer-Vogt-Sjögren-Batten disease and is a rare, fatal autosomal recessive neurodegenerative disorder that begins in childhood.

Vignette 2

Introducing James

James has just turned one year old. He is a typical one year old – sitting up, playing and interacting socially as one would expect! He is a little bit slow to crawl and walk but this is normal bearing in mind his history

Introducing His Family

James's mum, works for national healthcare charity. Her work experience has been very helpful as she understands support packages and commissioning. She has been on maternity leave but anticipates being back at work soon to flexible working.

James has a brother who is eight. It was particularly tough for his brother when James was in hospital, and bringing him home made an incredible difference to the whole family and especially to his brother. Throughout his brother was amazing – he has been unselfish and accepted they could not be together all the time.

James's family have been in touch with other families in similar circumstances, having met them on the ward or on Facebook. His mum also knew someone when she was pregnant who had a similar complication and she has become a good friend, particularly when their respective partners had to return to work.

Introducing Their Experience

- James spent his first eight months in hospital, the majority of which was in intensive care and had a long period of instability. He then had a short stint when he was medically stable, just six weeks, before he came home. He immediately improved after his heart operation and has continued to improve, and now does not need oxygen and purely has CPAP support to breathe. He is being weaned off the ventilator and is moving from four to five hours a day.
- James has a dedicated team who work together to ensure cover and are able to care for James when his mum goes back to work.
- Even though James was home relatively quickly a key challenge was overcoming silly and unnecessary obstacles. His mum observes “long term ventilated children are going to go home more frequently; the timescales in hospital are too long. The changes in James since he has been home are amazing. We need to make sure we get the children home more quickly – with no faffing”.

Understanding the condition

James was born with a diaphragmatic hernia which was complicated with heart problems, which ultimately caused the need for ventilation. His airways were compressed and has left a floppiness and a weakened airway. He has a drive to breathe and is expected to be off ventilation next year, with the trachy out in spring.

Vignette 3

Introducing Jenny

Jenny is five years old. She likes TV and loves going out and wants to go out every day. She has an on-going battle with her care team to go out every day which they now do with careful preparation and planning. Essentially she wants a normal life. She uses facial expressions and eye pointing to tell people what she wants and needs.

Introducing Her Family

Jenny's mum is an accountant and her dad was a sales engineer until his role was made redundant. His mum still works. As a family they have great friends and family locally, but whilst Jenny was in hospital, her parents lost some contact as they tried to spend as much time with Jenny as possible. Jenny now has a younger sister who is three.

Her mum has found real support from Facebook sites looking for advice of "silly things that no one else tells you". Her mum is looking to move to a personal budget because she does not want lots of individuals looking after Jenny – but one or two carers who know what she wants and will be great playmates. She comments "this will also give us flexibility for example to have someone look after Jenny while at after-school club."

Introducing Their Experience

- Jenny was born very small, and spent a week in neonates and then was discharged normally as a small baby. The family were concerned though as she seemed unwell, and was not feeding properly. Following a visit to their GP they went to A&E where her sats were low, she was grey and she went in to resuscitation. She was diagnosed with bronchiolitis and was about to go home when she suddenly stopped breathing. She went into ICU and was ventilated at 10 weeks. She made no respiratory effort and went through a whole barrage of tests – neurological, geneticand was eventually had a diagnosis of Spinal Muscular Atrophy with Respiratory Distress (SMARD). She continued to deteriorate and was down a palliative care route. But SMARD can occasionally plateau which is what happened with Jenny when at 8 months she stabilised and after 13 months in hospital came home.
- At two and a half she started at nursery and has just moved into reception. She remains completely dependent on the ventilator and is peg-fed but now has a weak swallow and can move one shoulder. The family are optimistic for the future and want to keep her lungs as healthy as possible.

Understanding the condition

SMARD is rare – there are 26 cases reported worldwide eight of which are in the UK. SMARD is a terminal neurodegenerative disease that affects the motor neurons and causes progressive muscle wasting (deterioration of feet, legs, hands, arms and neck muscles).

The predominating symptom is severe respiratory distress due to paralysis of the diaphragm. Most patients will experience respiratory failure within their first six months of life. Many of these children die in their sleep and most will not live past their first birthday. For those who do live, artificial ventilation is required. Progressive muscle weakness, especially affecting the lower limbs, follows the respiratory failure. Cognitive function is unaffected. Like all motor neuron diseases, there is no cure.

Vignette 4

Introducing Freya

Freya is six. She is a “feisty madam!” She attends mainstream school, and demands to be treated like a normal six year old. She wants to be able to do the same things as her sister and expects people to find a way. She particularly loves being on her tricycle as this gives her real independence. There are not many active fully ventilated children! She is partially deaf so uses British sign Language (BSL)

Introducing Her Family

Freya lives with her mum, dad, her sister who is seven and a half and brother who is two and a half. Her mum, was a Project Manager, but has not worked since the birth of her first child and plans to be at home until her youngest goes to school.

This has worked out well as her mum is not sure how she could juggle everything and manage the personal budget if she were also working full time.

Her parents observe that taking a trachy ventilated child into hospital for any appointment involves full parental involvement and clinical responsibility is expected to be taken by parents.

Introducing Their Experience

- Freya was born normally with no concerns. She has been home for six months when she became unwell with a chest infection. She then collapsed at home and went into her local hospital as an emergency where she stopped breathing. Having spent some time here she went on to Great Ormond Street Hospital where she was in intensive care for six months. She was trachy'd here and is now fully dependent 24 hours. Almost by accident they arrived at her diagnosis of Brown-Vialetto-Van Laere syndrome (BVVL) by trialling massive doses of riboflavin vitamin B12.
- Although usually degenerative Freya has shown improvement in all areas apart from breathing. When she first went into hospital she was sitting unaided, then she progressively lost all her movement, and then after 14 months started to move her hand. She can now independently sit, crawl, walk and tricycle!
- Her mum commented that “the step down from ICU to HDU was significant. The expectation of the ward staff was that the parents would stay and be the day shift – this was the worst part – the most upsetting time”. Before going home, Freya was moved to a children’s hospice which went well. Within weeks of being home she was perkier and happier.

Understanding the condition

The Brown-Vialetto-Van Laere syndrome is a rare neurological disorder which may present at all ages with sensorineural deafness, bulbar palsy and respiratory compromise. It affects the body's nervous system. BVVL is a motor neuron disorder: it attacks and progressively destroys motor neurons (the cells that control muscle activity including breathing, speaking, swallowing, and general movement of the body), causing paralysis of the cranial nerves. BVVL is usually degenerative. Recently it was demonstrated that in some patients the disease is caused by mutations in a gene which encodes the intestinal riboflavin transporter. In these patients riboflavin deficiency is the cause of the BVVL syndrome and supplementation of riboflavin proved a life saving treatment.

Vignette 5

Introducing David

David is almost three. He currently lives in a rehabilitation home. When he is well enough he goes to nursery every day and goes out on day trips with his family – which he loves. He also goes to the cinema every week. David likes singing and can do some signing. He does get tired quickly and fluctuates quite a lot – he can have some really good days and then some not so good days. David also has some wonderful carers and they do a great job with him but these are not carers for home.

Introducing His Family

David comes from a medical family, with both his parents working in healthcare. He has a sister who is four. She has severe asthma and has been in critical care herself. She has just started reception at school which involves over 20km travel every day. This is despite applying for the nearest school on social grounds. Both parents have significant health concerns that can affect their ability to care. None of the wider family health concerns have been considered in care package discussions. Neither has their wish or need to work been considered and they are frustrated as they feel that they are expected to give up work and become full time carers if they want to bring their child home. The stress of David's circumstances has put the family under huge pressure and his father no longer works full time as a result. His grandparents visit him regularly.

Introducing Their Experience

- David's birth was difficult at 34 weeks, with extensive bleeding. He was sent home and then had a cardiac arrest and was given CPR for 30 minutes by his mother. She has since been diagnosed with post traumatic distress disorder and is keen not to be placed in a situation where she is forced to resuscitate her child. Following his cardiac arrest at home he then had monthly respiratory arrests. He returned to hospital and has not lived at home since. Over time his condition has improved significantly but he is still not stable. The specialists suspect he has Brown Violette Van Laere syndrome (BVVL).
- He has had a single successful weekend visit home with a nurse. A weekend visit previously attempted with an HCA was blue lighted back to hospital. Having been back to hospital for a spell, he is just about to return to a children's hospital which caters for severely disabled children many of whom have particularly complex health needs. He is deemed ready for discharge but has instabilities that need addressing so until a safe and coherent care package produced, he cannot come home.
- Although initially paralysed David is making progress and can sit up on his own for an hour although he doesn't yet do it very often. They see daily improvements in him and are optimistic that this will continue in the long term.
- His parents have deep concerns though about the care package as this feels like an ongoing battle as inevitably his care is expensive and the commissioners have indicated they are no longer happy to fund his current placement (which David is thriving in) but until a care package for home is negotiated there is nowhere else for him to go. This puts an incredible strain on his parents and they feel "emotionally worn down". At times they feel there is no one to help though they have found the Breathe On charity supportive.

Understanding the condition

The Brown-Violetto-Van Laere syndrome is a rare neurological disorder which may present at all ages with sensorineural deafness, bulbar palsy and respiratory compromise. It affects the body's nervous system. BVVL is a motor neuron disorder: it attacks and progressively destroys motor neurons (the cells that control muscle activity including breathing, speaking, swallowing, and general movement of the body), causing paralysis of the cranial nerves. BVVL is usually degenerative. Recently it was demonstrated that in some patients the disease is caused by mutations in a gene which encodes the intestinal riboflavin transporter. In these patients riboflavin deficiency is the cause of the BVVL syndrome and supplementation of riboflavin proved a life saving treatment.

Vignette 6

Introducing Eva

When she was born the doctors only gave her only a few months to live as she was born with an extensive myelomeningocele.. “But Eva was determined to prove them wrong. Something she went on to do for the rest of her life.

Eva was an extraordinary girl. Someone who never failed to make you smile. Someone who got pleasure from your happiness. Totally unassuming. She never pre-judged anyone, ever.”

When she died Eva was 23. with a cognitive age of 10/11. At 23 she was very small for her age at just 33kg.

Introducing Her Family

Eva’s parents were both extensively involved in her care. She has a wider family, with sisters, nieces and nephews....all of whom loved being with Eva.

Some of her longer term carers became good friends of the family being in the home 12 hours a day 30 hours a week, and often knew more about her; her needs, likes and dislikes than the professionals.

Her family were clear that although understandably home needed to be safe and functional to meet her needs, it also needed to be a home not a hospital.

Introducing Their Experience

- Eva was born with an extensive myelomeningocele , this was not felt to be operable at birth and hence the open lesion on her back was not initially closed surgically and was only surgically closed when she was two years old. She had a shunt inserted into her brain at 9 weeks of age for subsequent hydrocephalus management. This worked well until she was 10 years old, it was then replaced and worked well until at age 17 years it began to fail.
- This was the start of major problems, she had 25 operations over the next four and half years and was hospitalised all of this time. Initially admitted with shunt problems, it was quickly apparent that the problems were caused by a chiari malformation (herniation of the brain); following the second operation to cure this Eva succumbed to a life threatening infection which caused her to lose use of her arms and she was from this time unable to breathe independently and required 24 hour ventilation. Up until this time we had never required any carers to help to look after Eva but with the introduction of long term ventilation this all changed.
- The problem was that she was from 17 - 21 in a paediatric hospital on HDU, when she was discharged home, there was no equivalent in adult health care to the holistic approach used in paediatrics. In paediatrics all specialists are housed under one roof. When discharged home Eva was under the care of 8 adult hospitals for different body systems ie ENT etc but none were able to take on her long term ventilation needs, none were used to involving parents / carers in consultation appointments. All found her to be "Too small" "Too complex" for their services. The local GP would not even see her as she was "beyond his level of expertise".
- After 10 months at home with no adult hospital / Dr ; in charge of Eva's long term ventilation , we as her parents emailed a long term ventilation unit in London. They agreed to see Eva for a review and at the first review accepted Eva as their primary patient. For the next 18 months our health provider for Eva was this unit in London and we lived in North Wales.

Understanding the condition

Myelomeningocele is a birth defect in which the backbone and spinal canal do not close before birth. The condition is a type of spina bifida.

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