A Review of the Benefits, Challenges and the Future for Interfaces for Long Term Non-Invasive Ventilation in Children

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Abstract

Long Term Non-Invasive Ventilation (NIV) is increasingly common and is benefitting children with a wider range of conditions. It improves quality of life and life expectancy and reduces hospital admissions and length of stay. Children are at risk however of adverse effects caused by NIV interfaces such as skin injury, facial flattening and eye problems. The correct size interface, that is properly fitted, can decrease the risk of device related injury but it remains difficult to fit interfaces to children with particularly small, syndromic or asymmetrical face shapes. Specialist centres are producing semi-custom and fully custom-made interfaces in an attempt to improve fit and decrease device related injury. Recent advances in modern technology such as facial scanning and 3D printing are making custom made interfaces a more viable solution, but it is not yet known whether these approaches will be cost-effective and what the impact of these interfaces will be on the adverse effects of NIV.

Benefits of NIV

There is evidence that the use of NIV both improves the quality of life and life expectancy of children with neuromuscular disorders. NIV is effective in correcting nocturnal hypoventilation and in alleviating the associated symptoms [5-9]. The evidence clearly shows that those who decline NIV are more likely to die of respiratory failure within 2 years [10-16]. NIV has been shown to reduce both hospital admissions (2-4 per year pre-NIV and 1 per year post-NIV) and days spent in hospital (40-50 per year pre-NIV and 10 per year post-NIV).

Introduction

Ventilation delivered through the patient’s upper airway using a mask or comparable device is termed Non-Invasive Ventilation (NIV) [1]. NIV was developed in the 1980s to support breathing in patients with neuromuscular weakness [2,3]. More recently, the use of NIV has been offered to an extended group of patients to treat nocturnal hypoventilation, central and obstructive sleep apnoea. Untreated nocturnal hypoventilation leads to daytime hypercapnia which causes headaches and drowsiness. Untreated obstructive episodes lead to sleep fragmentation with daytime somnolence as well as chronic hypoxia resulting in pulmonary hypertension and cor pulmonale [4].

Keywords

Children, Long term non-invasive ventilation, Interface

Cerebral palsy
Neuromuscular
craniofacial problems
airway malacia
posterior fossa haemorrhage
Central apnoeas
CCHS

Figure 1: The seven most common underlying conditions for children using NIV in a UK specialist centre.
[9,17,18]. Further work to look at this is underway within our group [19].

There has been a significant increase in the use of NIV over recent years with census surveys showing a rise from 71 in 1998 to 705 in 2008 [20,21]. The reasons for this include increase in life expectancy of preterm babies, increase in conditions such as obesity and increase in public expectation and demand. A more recent survey completed by our group in a UK specialist centre indicated the diversity of use of NIV in children and the conditions it is predominantly used for (Figure 1).

Evidence indicates that Sudden Infant Death Syndrome (SIDS) is associated with specific pathological abnormalities in the medulla involving neuromodulators known to be involved in the neuronal control of breathing [22]. Hence, in infants that demonstrate significant disturbance of respiratory control, NIV is often used as a preventative measure. Yet, there is no evidence that this therapy improves outcomes, and, as disorders of respiratory control often improve with maturity, NIV can be withdrawn when gaseous exchange parameters improve or resolve.

Although NIV has become increasingly effective and popular, the success of NIV is primarily dependent on the interface between the ventilator and the patient rather than any other aspect of the intervention e.g. mode of ventilation [23,24].

Interfaces

The main role of the interface is to create a seal with the patient’s face allowing the delivery of air pressure needed for effective ventilation. There are broadly six different types of interfaces available for the delivery of NIV (Figure 2a, Figure 2b, Figure 2c, Figure 2d, Figure 2e and Figure 2f). These are mouthpiece, nasal pillows, nasal mask, oronasal/full face mask, total face mask and hood.

The advantages and disadvantages of these different types of interface have been described fully by various authors for many years and factors include comfort level, claustrophobia, aspiration risk and hypersalivation [25,26]. The net result of these factors however is that nasal and oronasal masks are the most commonly used interface [26,27]. Nasal masks are deemed to be the most comfortable but nasal and oronasal masks are equally effective in terms of the ventilation delivered [28]. Interfaces are selected for individual children based on a complex multifactorial evaluation of clinical condition, face shape and size, child/family preference and professional experience/expertise. Sizing guides are provided by manufacturers to aid in this decision making once a type of interface has been chosen but it is still common for multiple interfaces to be tried.

Adverse Effects of Interfaces

Whilst significant research and development has gone into the design and production of NIV interfaces,
interface related adverse effects are still widespread and ‘medical devices are the leading cause of paediatric pressure ulcers’ [29]. Air leaks around the interface are frequently the reason for NIV being ineffective and/or poorly tolerated. Although air leaks can be minimised by careful interface selection, efforts to minimise or stop air leakage are the most common cause of interface related adverse effects (Figure 3). Frequent alarming of the ventilator, particularly at night, causes sleep disturbance and high levels of parental anxiety.

These adverse effects can be divided into three categories [25,26,30-32].

1. Skin injury
2. Facial flattening
3. Eye problems

Skin injury is caused by either pressure or friction at the site of mask contact [31,33]. High pressures on the skin from the interface are caused by high tension in the interface straps whereas friction results from movement of the interface, e.g. from when a person changes body position during sleep leading to a lateral shift of the interface or from variable tension on the ventilator tubing changing the position of the interface. The level of skin injury ranges from transient erythema, prolonged erythema and, in the most severe cases, skin necrosis. In its mildest form, skin injury is cosmetically unsightly with red areas of skin, but in the more severe situations poses a health risk as it may become impossible to ventilate the person non-invasively necessitating an invasive interface, e.g. intubation or tracheostomy.

The type of interface used dictates the areas of skin most likely to be affected but skin damage is most often seen on the bridge of the nose [29,34] (Figure 4a) or forehead but the sides of the nose can also be affected (Figure 4b). Efforts are made to prevent or reduce damage using pressure relief dressings which can improve the situation but doesn’t address the underlying problem [29,35]. Another approach is to alternate the type of interface chosen to vary the area of skin insulted but this is also a short-term solution.

Facial flattening is a problem particularly experienced by the paediatric population. Pressure from an NIV interface on the growing face can result in under-development of the maxilla, leading to mid-face flattening and malocclusion of the teeth (Figure 4c). Depending on the choice of interface and the consequent areas of pressure, facial changes may be global or restricted to specific facial regions. Whilst considered to be a long term adverse effect, these changes can occur after as little as 4 weeks use of NIV [30] and are essentially irreversible without significant surgical intervention. The success of NIV at increasing life expectancy means that many of these children will now live for a number of years with this facial deformity.

Hoods have been proposed as a solution to both skin injury and facial flattening as they remove the need for pressure on the skin and bones of the face and some success has been seen in the care of adults [36]. They do however transfer the pressure areas to other parts of the body such as under the arms and potentially around
the neck. Hoods also pose other problems as the higher volume in the hood compared to other types of interface can lead to poorer ventilator interaction due to delayed triggering [37].

Eye problems occur either as a direct result of air leaking from an interface into the eyes or where the eyes are within the interface, such as a total face mask or hood. In both cases, the air is usually cold, dry and under pressure, resulting in eye irritation and/or dryness. Eye irritation is less common than other side effects [33,34,38] but still contributes to NIV failure. The problem is addressed by adjusting the fit of the interface (by altering the strap tensions etc) or by changing to a better fitting interface.

Headgear

NIV interfaces are usually held in place using commercially produced headgear. These take the form of fabric straps or a cap with straps attached. The headgear can, in itself, be a cause of interface related problems [39] where the pull from straps is not in the ideal direction to seat the interface with even pressure on the face or the straps provide discomfort where they cross the tops of the ears or face prior to reaching the interface. The tendency is to over tighten straps in an attempt to reduce air leaks, but this results in increased pressure on at risk areas of the face, such as the bridge of the nose, leading to skin damage [40]. Education is important for both professionals and patients/families to ensure that, whatever type of interface-headgear combination is being used, it is fitted in the most appropriate manner with the minimal strap tension possible used to achieve the best fit.

Interface Fit

For NIV to be used successfully, the fit of the interface is critical. Although a variety of interfaces are available commercially for standard-size adults and children, the same range is not available for small infants and a good fit is difficult to obtain in syndromic children and those with neuromuscular disease or other types of facial deformity, for example craniosynostoses [30,31]. It is in these groups of children however that NIV may be most beneficial.

The current system for fitting interfaces relies heavily on the experience of the clinician in visually assessing the child’s facial features accompanied by a degree of trial and error once the choice of interface has been narrowed down. Sizing guides are provided by manufacturers to aid in this decision making but it is still common for multiple interfaces to be tried. The process is further complicated by continual growth of the child’s facial structures and that the pattern of growth may be affected by the application of the interface itself.

Recognition of the pivotal role the interface plays has driven the technical development of interfaces to try to overcome the above problems. Each interface type is available in varying shapes, sizes and construction materials and the range has been further increased by the advent of semi-custom-made interfaces in addition to the standard commercial offerings.

Semi-Custom-Made Interfaces

Semi-custom-made interfaces are bought ‘off the shell’ and then customised to fit the individual. The method of customisation varies depending on the individual design and options include a thermoformable plastic frame, a fixed frame in which quick drying filler is injected [26], an inflatable mask cushion or a silicone gel filled cushion which moulds when heated [25]. It would be expected that semi-custom-made interfaces would offer an improved fit in comparison with standard interfaces, but no studies have been performed to make this comparison. Although semi-custom-made interfaces are offered by a number of manufacturers, clinical experience indicates that they have not seen widespread adoption indicating that they do not offer significant benefits over standard interfaces.

An alternative form of semi-customisation involves the mixing and matching of different interfaces and headgear to achieve the best possible fit with equipment that is available. This is often done by clinicians as part of the fitting process, but it is known for more extreme measures to be taken by parents, for example, stitching headgear to make it fit or removing interface parts which cause areas of pressure damage to the skin.

Custom Made Interfaces

The lack of adequate interface solutions has prompted a few centres specialising in NIV to create custom made interfaces for their patients. In this situation, a personalised mask is made for an individual patient. The methods for making custom made interfaces varies significantly and includes the use of thermoformable plastic modelled on plaster phantoms and then individually adjusted [31,32] as well as acrylic masks created from a plaster model of the face (made from an individual alginate impression) [41]. Technological developments now also enable non-invasive 3D modelling of the face using facial scanning techniques such as structured light [42], although this technique appears to have been used primarily to date for the construction of partially custom-made interfaces [43]. The method adopted, however, has depended on the expertise available locally and techniques remain centre specific without widespread dissemination to other centres or groups of patients.

A limited number of studies have been carried out investigating the performance of custom made interfaces in comparison to the standard alternatives, both for paediatrics and adults. Overall, Custom made interfaces are well tolerated and comfortable [32]. In adults, they have been shown to improve ventilation [32,41] and, in children, custom made interfaces are associated with a
lower incidence of skin injury [30]. No cases of eye irritation have been reported whilst using a custom-made interface [32] but custom-made interfaces have yet to show a beneficial effect on facial flattening [30]. However, in no studies have custom made interfaces been found to be inferior in comparison to commercially produced interfaces.

Cost and Time

The studies performed to date demonstrate that custom made interfaces are at least equivalent to, and in most cases better than, standard commercially produced interfaces. Very little information is available however about the time and cost involved in producing these personalised interfaces. It is thought to take up to three hours to make an interface, depending on the method used, and in 1999 cost US$90 for an interface that would last approximately 18 months [41]. The average cost for a commercially made paediatric nasal mask is currently approximately £60 and more recent figures indicate that the initial costs for a custom-made interface are higher than a standard product, but the custom-made interface is more cost effective in the longer term (ie at 5 years) [44]. When a cost comparison is made it is important to take into account the number of interfaces that may be ‘wasted’ through the trial and error process of finding the commercial interface that provides the best fit. The economic evaluation is, however, far more complicated than a direct comparison of interface costs. In our experience, adverse effects from poor interface fit lead to increased numbers of home visits, clinic appointments and, in the more severe situations, admission to hospital. These events all carry a significant cost, both financially and in terms of time and quality of life. Prevention is undoubtedly better than cure and a well-fitting interface would render these events avoidable.

Additional Design Considerations

Although the fit of an interface is of primary importance, when manufacturing interfaces it is important to take into account other considerations such as dead space and the impact this has on CO₂ rebreathing. CO₂ rebreathing is affected by interface volume, flow during expiration and position of an exhalation port within the interface [45,46]. Interfaces also differ in design depending on whether they are vented or non-vented i.e. presence or absence of an anti-asphyxia valve and an exhalation port [25]. All these aspects need to be taken into consideration when designing new or custom-made interfaces.

The Future of the NIV Interface

Technological developments offer the opportunity to improve on the traditional methods used for the design and construction of custom made interfaces. It should no longer be necessary to use a physical facial model (either from a phantom or the patient) to create the form of the interface. Non-contact alternatives would render the process much quicker and significantly improve the patient experience. Electronic transfer of the patient’s facial data would then enable rapid automated production of the interface cutting down the time from assessment for the interface to the patient receiving their custom-made interface. Custom made interfaces produced through improved methodologies should lead to improved fit. It is an assumption however that they will provide an improved fit and that improving the fit will lead to further reductions in interface related adverse effects. Robust research will be required to investigate this new generation of custom made interfaces with specific regard to comfort, leak, skin damage, facial deformity, eye irritation and economic evaluation.

Conclusion

NIV is effective but relies on a good user-ventilator interface for optimal ventilation and user compliance. Achieving a good interface fit is difficult and particularly so in small children and those with atypical or asymmetrical facial architecture. The range of commercially produced interfaces is now quite significant, and the advent of sizing guides aids the choice of an appropriate interface, but adequately fitting interfaces are not available for all patient groups. Some specialist teams have created custom made interfaces which have been shown to reduce the adverse effects associated with NIV but they are difficult to produce, expensive and are not widely available. Modern technologies offer the opportunity to develop systems for custom made interfaces to be produced quickly and economically, however, research is needed to demonstrate whether this new generation of custom made interfaces can further address the adverse effects currently associated with NIV.

Educational Aims

- Understand the primary groups of children using long term non-invasive ventilation and the benefits afforded by it.
- Understand the types of interfaces available and the impact of interface related adverse effects caused by long term NIV use.
- Understand the importance of correct interface size and fit.
- Be aware of current and potential future developments in the provision of custom made interfaces.

Future Research Directions

Future research should be directed towards ameliorating the adverse effects of long term non-invasive ventilation. Development of an algorithm to improve successful selection and fitting of commercially available interfaces may reduce the reliance on the expert clinician and enable optimal fit to be achieved for more children. For those where a good fit is not achievable.
with an off the shelf product, clinical studies need to be carried out to investigate whether custom made interfaces do reduce interface related adverse effects. Work will also be required to evaluate the economic impact of custom made interfaces for non-invasive ventilation.

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References


