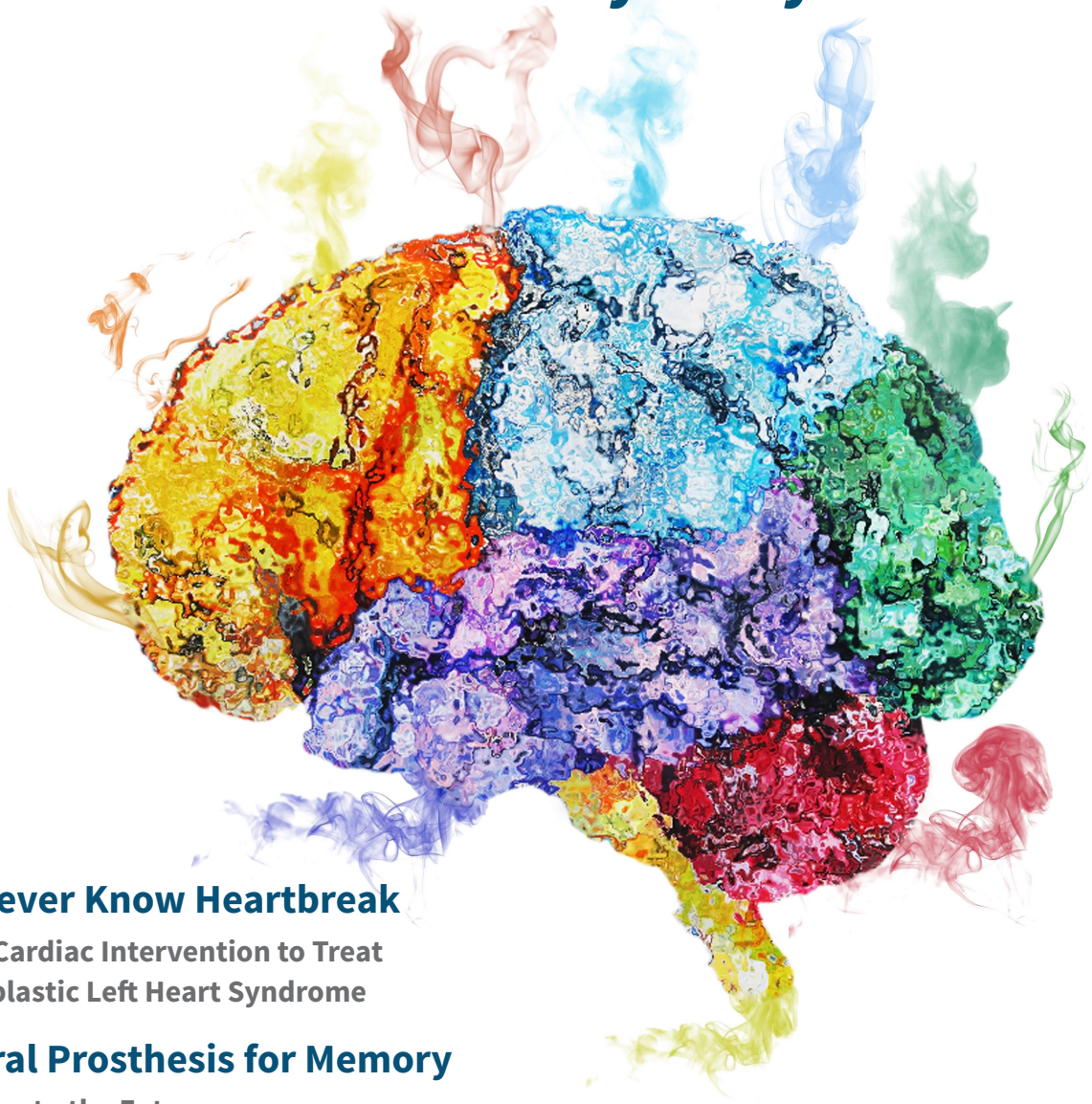
TSMJ
Trinity Student Medical Journal

Neurosurgical Treatment
of Moyamoya Disease



To Never Know Heartbreak
Fetal Cardiac Intervention to Treat Hypoplastic Left Heart Syndrome

Neural Prosthesis for Memory
Looking to the Future

Irish Doctor Exodus
Why the Irish Health System Cannot Retain Its Junior Doctors

The *Trinity Student Medical Journal* is intended to provide an inclusive vehicle for students to communicate current medical research, opinions and thoughts to other students, faculty members and faculty of affiliated hospitals and institutions. We publish articles relating to many aspects of medicine including scientific research and clinical experience. Articles are accepted from students in medicine and other related fields, as it is our view that medicine is the meeting point of any disciplines. The aim of the Journal is to provide a medium that is responsive to the rapidly changing face of contemporary medicine, and is able to grow and expand as rapidly as the subject.

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Abbreviations

TCD: Trinity College, Dublin
RCSI: The Royal College Of Surgeons, Ireland
UCC: University College, Cork

TSMJ

Director's Welcome

Dear Reader,

As director, it is my pleasure to welcome you to the 14th edition of the *Trinity Student Medical Journal*. For those unaware, the Trinity Student Medical Journal is written, edited, and produced by medical and health sciences students, and is the result of months of work from both the committee members and the students/faculty who have submitted their work. It is a means for talented and devoted students to share their interests. And the papers selected for this edition, we believe, contained the most significance of what the *TSMJ* strives for. Chosen or not, I would like to thank all those who submitted articles as we recognise the hard work and commitment you all have provided. And we hope that the articles contained within will inspire students to continue their interests in research.

As director this was a new experience for me, as in the past I had dealt mainly with production. This would be my third year with the TSMJ, and I have seen the journal progress significantly during my time. It has been a big learning experience and I have learnt even more than I ever thought I would about the TSMJ and the journal form itself. It amazes me how we are able to improve and build upon previous editions in our quest for an accomplished platform for students with such little time. This year we acquired a website domain of our own as a means to provide even easier access to the journal and its articles. These years with the TSMJ have been a great experience and I am proud to have seen the journal thrive and continue to grow.

I would like now to take the time to thank everyone in the committee for all the hard work they have provided in keeping the journal at a high standard.

To our editors-in-chief, Orna Grant and Conor Lavelle, who worked unremittingly as both leaders and editors; thank you for supporting me and helping me with my role as director even though I had lacked the experience.

To all of the editors, thank you for your tireless hard work and contributions. They may often go unnoticed, but they are indeed crucial to the achievement of the journal. It is your work that maintains the utmost quality to which the TSMJ strives.

To our marketing team members, Abdel Satt Rubayawi and Albert Kelly, thank you for spreading awareness of the journal and the commitment to obtain its adequate resources. Without your talent we would not have the means to provide a journal.

To our Graphics designer, Niall Byrne, thank you for your amazing, and original artwork that graces the pages of the TSMJ. It shows that the journal has expanded and matured to include original work directly based on articles.

And to our Production team, you have done an unbelievable job. Jaclyn Vertes, David Parfrey, and Stephen Sheridan, you are all an amazing team to work with, and I would like to thank you all for allowing the journal to materialize. Ironically much of my experience with the TSMJ was based through production, and yet you all required little leadership or guidance from me. In fact you all managed to improve the journal even further. Congratulations on a work well done.

I would like to give a special thanks to Antonio Bueno, the previous Director of the TSMJ, for all the advice and mentoring you provided me with. Without your help and knowledge from past experience I would've been lost. Thank you for the guidance and making me strive to be a better leader.

I wish a great, big thanks to all of our sponsors and supporters. Without your generous contributions we would not be able maintain and produce the journal. Thank you for continuing to provide a platform for students to be heard and interests shared.

Lastly, I would like to thank you, the reader, for providing us your interest and allowing us to share the work we have put into the journal these past few months. We hope that you enjoy this edition and that it may inspire you to contribute or be a part of the Trinity Student Medical Journal in the future. Thank you.

Sincerely

Peter Tsakkos

Director
TSMJ 2013

Editorial

Dear Reader,

Some of its detractors maintain that scientific writing does not possess the same level of artistry or beauty as literature, poetry, or fictional prose. For many of us, that these detractors cannot appreciate the depth of meaning and artful design of science writing is akin to missing the genius of Yeats or Kafka. Scientific literature is capable of attaining the same artistic heights as the finest short story, the most compelling play, or a well-spoken verse of poetry. Researchers and science writers help to unearth incredible facts about the way the universe conducts itself, and their work often has a profound impact on our relationship with the world in which we live. In a sense, scientific writing is the culmination of human curiosity and artistry, and we believe that it is something to be celebrated.

Each year, many hours of hard work go into the production of the *Trinity Student Medical Journal*, and this year has been no exception. The tireless efforts of our editing, marketing, and production teams are of particular note, and we would like to extend our gratitude to all those involved.

We invite you to explore this year's *Trinity Student Medical Journal* with the same sense of discovery and curiosity that went into its production.

Sincerely

Conor Lavelle and Orna Grant

Editors-in-Chief
TSMJ 2013

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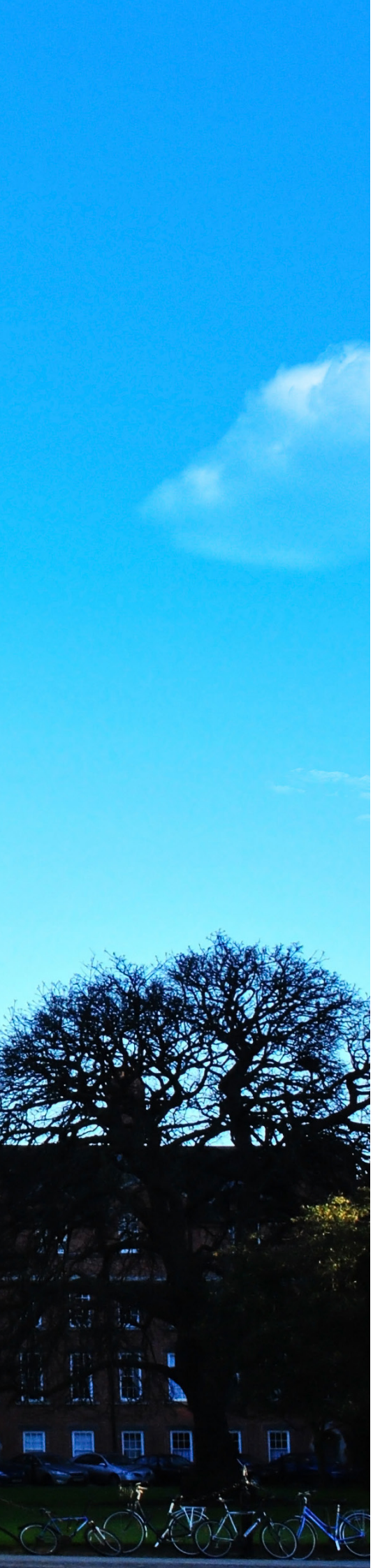

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Eoin Kelleher, 4th Year Medicine, RCSI

> "Is it any wonder that most of our NCHDs are leaving, and many of those who remain regret entering the profession?"

Introduction

Well-trained and motivated medical staff are essential to a functioning health system. However, Ireland is struggling in this regard. Ireland is the EU country with the highest proportion of its doctors working abroad[1], with 47% of our medics working outside the country. For example, In 2011, half of all graduates left Ireland after intern year[2]. Malta is a distant second with 23.1% of their doctors leaving. As has been highlighted in numerous studies and reports over the years, Irish non-consultant hospital doctors (NCHDs) – those doctors in training and other temporary posts – are demoralised and dissatisfied with working conditions in Ireland and are leaving[2-9]. Half of NCHDs report being dissatisfied with their current job in Irish hospitals[3,4], and 57% would not recommend a job as an NCHD to a member of their family[4]. In addition, 32% told the Irish Medical Organisation (IMO) Benchmark Survey in 2011 that they would not choose medicine again if they had a choice[4]. This backs up findings from the Career Tracking Survey (CTS) of 2005, which surveyed Irish doctors who graduated in 1994 and 1999. This survey found that only 70% of graduates would train as a doctor again[9]. These figures compare poorly with corresponding information for doctors working for the National Health Service (NHS) in the United Kingdom[10-12]: in 2012, 82% of graduates from 2006 in the UK had a strong or very strong desire to practise medicine, and fewer than 1% regretted becoming a doctor[10].

A systematic review by Willis-Shattuck and colleagues explored reasons for health-worker retention in developing countries and identified seven motivational themes[13]. This article reports the views and experiences of Irish NCHDs under these themes, using information derived from recent reports and studies.

The seven themes identified by Willis-Shattuck are:

1. Financial incentives
2. Career development
3. Continuing education
4. Hospital infrastructure
5. Resource availability
6. Hospital management
7. Personal recognition or appreciation[13]

Financial Incentives

Although financial incentives are important, they are not sufficient to determine the retention or emigration intentions of doctors. Rather, monetary rewards are one of many factors which affect physician morale and motivation[13,14]. A 2012 survey of Irish NCHDs found that over half were dissatisfied with their pay, but less than a third reported it as an important factor in any decision to move abroad[3]. An important issue affecting NCHDs is widespread breaches of their contract. Non-payment of unrostered overtime by hospitals has been widespread in recent years, with 55% reporting to the Irish Medical Organisation (IMO) that they do not get paid for all the hours that they work[4]. This is widely cited as a major cause of upset in surveys of NCHDs and in the media[8,15]. The withholding of pay often leaves workers feeling demoralised and undervalued by the health service for which they work, and contributes to negative attitudes towards hospital management which will be further outlined below. Several cases have been taken by the IMO to the Labour Court, which is the last resort for industrial relations disputes[16].

Career Development

Defined career development opportunities abroad are identified as important factors in deciding to emigrate[13]. Only 16.4% of NCHDs surveyed in 2012 thought their chances of obtaining a consultant post in Ireland were "good" or "excellent", while almost half thought their chances were "poor"[3]. These findings echo the Career Tracking Survey (CTS) which found that the most important factor in encouraging Irish doctors to return to Ireland was the availability of consultant posts[9]. Numerous reports have highlighted the need to move towards a consultant-delivered health care system with an increased ratio of consultants to NCHDs, notably the Tierney Report, Hanly Report and Buttimer Report[17,18,5]. In clinical directorates, consultants and NCHDs work in teams to provide care, rather than each consultant post being supported by a team of NCHDs. However, despite the many reports, there has been limited progress on this. Many NCHDs remain in registrar posts even once they have completed their training, partly because the base of the pyramid is too wide and the ratio of NCHDs to consultants too high.

Continuing Education

Training is important for doctors because it allows them to develop professionally and achieve personal goals[13]. The poor quality of training available in Irish hospitals is consistently highlighted by NCHDs as a problem, and seeking better training is often given as the main reason for leaving Ireland[3,8,9]. In the 2012 survey, 40% of NCHDs rated the training they received as "poor"[3]. A significant proportion of respondents to the CTS in 2005 still working in Ireland reported "poor structure, quality and organisation of training" as a major problem (19% of the 1994 cohort; 25% of the 1999 cohort)[9]. However, there was a wide variation across specialities, with over half of graduates rating training as a major concern in medical specialities[9].

Here's a postcard from your doctor, he says he'll be a bit late.

The Second Interim Implementation Report of the Reform of Intern Year of the Health Service Executive Medical Education and Training committee (HSE-MET) showed that 44% of interns felt the training provided was "poor"[2]. 77% of respondents received less than three hours of formal teaching per week. A "no-bleep" policy during teaching was adhered to in only 5% of cases[2]. 94% experienced difficulty being released from clinical duties to attend training sessions[2]. 25% never received feedback from their trainers, and 62% only received feedback "sometimes"[2]. The majority of NCHDs report that most of their training comes from informal and "on-the-job" sources rather than formal, direct training[4].

Much of NCHDs' time is spent on non-medical duties, such as "administrative tasks, organizing tests, finding charts etc." Over two-fifths of graduates from 1999 in the CTS report spending time on inappropriate tasks as a major problem[9]. 55% of interns report spending 60-80% of their time performing non-medical tasks, while 25% report spending 80-100% of their time[2]. These findings are supported by the IMO Benchmark Survey[4].

While it is generally accepted that doctors carry out some further training abroad to gain valuable experience to bring home to Ireland, it is clear that Irish medicine needs a culture change. We must provide an environment in which the role of NCHDs is no longer seen as primarily service-provision, but as a training post where young doctors spend their time learning so that they may practise medicine as part of teams of consultants. This suggestion has been advocated before, most notably in the Buttimer Report[5].

Hospital Infrastructure and Resource Availability

Poor infrastructure and working conditions drain staff morale and affect performance and patient care. In the 2005 CTS survey, 15.5% and 13.5% of 1994 and 1999 graduates, respectively, reported working conditions to be a major issue[9].

Hospital Management

Skilled managers have the ability to motivate their healthcare workers, to advocate on their behalf and to respond to their concerns, resulting in a motivated and effective workforce[13]. Lack of support from management is commonly cited as a major problem working in Irish hospitals[8,9]. The widespread non-adherence to the European Working Time Directive (EWTD) and reported non-payment of unrostered overtime are two examples[4].

The EWTD mandates that healthcare workers cannot work more than 48 hours per week[19]. It was introduced in 2004, and was to be fully implemented by 1st August, 2009[9]. In 2011, only one-third of NCHDs were compliant with the EWTD, according to the HSE[9]. Over three-quarters of NCHDs report that the EWTD has not been implemented as of 2012[3]. Working hours are cited across all surveys as a major reason for leaving Ireland for countries with perceived better working conditions[2,3,8,9].

Personal Recognition or Appreciation

Recognition of the value of one's work by employers and the community was found to be an important motivating factor for healthcare workers[13]. Irish NCHDs, however, feel under-appreciated and demotivated by the health service for which they work. Evidence of extremely low morale was picked up in the IMO Benchmark Survey of NCHDs: as mentioned above, 57% would not recommend a career as an NCHD to a family member and almost one-third would not choose medicine again[4].

Consequences

Ireland makes up for the shortfall in Irish NCHDs by actively recruiting doctors from abroad, often from developing countries such as India, Pakistan and Sudan. There are issues with this practice, both practical and ethical. It is costly to recruit doctors from overseas, particularly when many will only stay for a few years. Moreover, there are ethical problems when a health service actively recruits medics from developing countries and thus deprives people there of much-needed medical

staff. This article does not attempt to address this other side of the issue; however, an article from RCSI and Trinity College, Dublin examines it in detail[20].

Conclusion

The above findings, which represent a serious threat to future medical workforce sustainability, should not be a surprise, considering how little attention is paid to the needs of NCHDs in Ireland. Training is perceived to be of poorer quality than in other countries to which Irish medical graduates can easily emigrate and its importance is not emphasised or recognised. In recent years, training allowances and grants have been cut, NCHDs are expected to work long hours that breach the EWTD and NCHDs are regularly not paid for this extra time. In addition, these long hours negatively affect their performance and ultimately put patient care at risk[21]. Furthermore, much of their time is spent performing non-medical tasks. Between all of this and widespread breaches of the NCHD 2010 contract[4], is it any wonder that most of our NCHDs are leaving, and many of those who remain regret entering the profession?

It says here that she leaves behind a husband, 3 children, 549 Facebook friends and 356 Twitter followers. Terrible sad.

This article makes clear that there is no easy fix, as there is no one reason driving NCHD emigration from Ireland. Future work will require a systematic analysis looking at how each of the above issues contributes to emigration, and ways to address it. New graduates and current NCHDs are – for better or for worse – the future of Irish health care. The importance of retaining them and motivating them cannot be overstated.

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Neural Prostheses for Restoring Memory – Looking to the Future

Seán Maguire, 4th Year Medicine, TCD

> "Restoring, enhancing or preventing loss of memory are tangible and realistic goals for the future."

Abstract

Neural implants are a new tool in the field of medicine, and are becoming increasingly common. Issues with memory are one potential area in which such devices may prove useful. This paper reviews research published to date and explores future possibilities and hurdles in developing a device available to the general population.

Introduction

Memory loss and deficits have long been a major concern among all societies. Traditionally associated with the elderly, conditions such as neural atrophy, Alzheimer's disease and other forms of dementia not only deprive individuals of cherished memories, but may also remove their independence and reduce capacity to fully function for the rest of their lives. Memory is poorly understood on a biological level, but new ground has been broken and some inroads have been made into understanding its inner workings. Utilising recent advances in our understanding of memory, hypothetical targets and possible neural interfaces have been put forward. Most have yet to see the light of day, but the door has certainly been opened, and restoring, enhancing, or preventing loss of memory are tangible and realistic goals for the future.

Biology

Though well researched in the field of psychology, memory in a purely biological sense is less well understood. Several types of memory exist, which tend to be concentrated in specific parts of the brain. Though not yet conclusively proven, there is a general consensus as to the general principles of memory formation and function.

Implicit procedural memory, i.e. memory of skills and tasks, is retained in the cerebellum at a largely subconscious level[1]. For example, an experienced driver can hold a conversation while driving, but a learner may find this overwhelming as they are

dedicating a higher proportion of conscious effort to controlling the vehicle.

Short-term/working memory is concentrated in the parietal and prefrontal cortices[2].

Memories in the traditional sense, as in recalled past experiences and emotions that are stored on a medium to long-term basis, are stored in different locations. Sensory information passes through the hippocampus to the medial temporal lobe. Medium-term memories are stored in the entorhinal, perirhinal, and parahippocampal cortices, whereas longer-term memories are moved to the neocortex, usually during sleep.

Difficulties with memory can affect any or all of the above areas, with correspondingly varying effects. Memory loss is generally broken down into two distinct types: retrograde amnesia, where one cannot retrieve past memories, and anterograde amnesia, where new memories are hard to lay down. The latter is usually trauma-related, but the former can occur not only from trauma, but a wide range of other causes such as Alzheimer's, alcoholism or simply old age.

Memories are laid down using a neurological activation pattern known as long-term potentiation (LTP), with a required growth factor, brain-derived neurotrophic factor (BDNF), which encourages consolidation of new synapses.

Memory retrieval occurs via action potentials (APs) that travel from the prefrontal cortex (where the decision to retrieve information is made), to the basal ganglia and from there to the relevant areas, as outlined above. The former two areas could be considered "gatekeepers" to the memories, so damage here would be devastating, but the memories are likely to remain intact in other areas.

Using this principle, we can consider lost memories not as gone, but currently irretrievable, as the storage locations are theoretically intact. This echoes the description of dementia according to Penny Garner of the Contented Dementia Trust; she describes memory as an ever-expanding photo album where new memories ("photographs") are not always inserted. Similarly, one could describe Alzheimer's as losing specific photographs throughout this album; the patient may be able to tell that there is an empty space in the album, which can cause considerable distress for sufferers[3].

Existing Research

Little research on actual devices has been done to date, though a single piece of literature has put forward a prototype.

This study looked at using a 32-electrode device implanted into the hippocampus of the rat brain. This was placed in the CA1 and CA3 regions, which connect up predominantly to the entorhinal cortex, which controls medium-term memory[4].

Following surgical implantation of the device and a minimum of 7 days' respite, rats were trained using a presentation of two levers. The inappropriate one had been previously presented, and selecting the other resulted in a reward, after which both levers were withdrawn until the rat had moved away for some time.

The implanted device recorded the brain signals in these areas via a computer connection. Using this data, characteristic brain signal patterns were recognised for each decision of the rat. A multi-input/multi-output (MIMO) model of electrical signals was used in this study to characterise the "memories" made during the training. The researchers had previously developed this model for such a purpose in mimicking hippocampal signals[5].

Thus, using this measured pattern as a template, the researchers could accurately predict the rats' choice from the brain output signal.

This MIMO signal pattern was then replicated and delivered to the rats. This had the effect of increasing the success rate of the rat selecting the appropriate lever. This improvement was

particularly evident when there was a longer delay between successive presentations of the lever.

A neurological agent, MK801, which significantly impairs neurological transmission within the hippocampus, was also used to simulate an impaired ability to remember. It produced a clearer result in the difference between stimulation and none.

The animals themselves acted as their own controls; stimulated and unstimulated success rates were compared among the same animals.

Significantly, the researchers also delivered a scrambled signal from the MIMO signals. This had no effect on success rate compared to no stimulation (control) success rates. This would suggest that the pattern of signal was important in influencing behaviour.

Implementation

In the context of Alzheimer's or another dementia, this approach may be of limited practical potential. This system requires signal measurement of the brain in advance of memory loss, i.e. "backing up" specific memories.

However, for non-patient-specific memories, such as procedural memory, this approach could be useful. Visual/emotional memories are unique and thus not retrievable without prior recording, but skills such as walking, speaking and bladder control are all learnt neurological responses, usually in areas of the cerebellum. If this approach were taken, a "stock" of skills and procedural memory tasks could be drawn upon to restore basic skills to individuals who may have lost them.

However, this brings up ethical concerns akin to those raised by gene therapy or gene selection. Theoretically, if such a system were to exist, one could essentially "shop" for new skills such as another language, ability to play an instrument, sports skills, etc.

Developing a neural prosthesis for memory is not an easy task, the brain being a highly complex and compact structure. Currently, we know roughly where much of it is stored anatomically, but the underlying mechanisms for memory formation and retrieval are still largely elusive.

Memory is not a uniform entity either, and great variations exist among individuals. One famous example is that of a lady known as AJ, who was one of the first documented cases of highly superior autobiographical memory[6]. She had an innate ability to remember details, banal and emotional, happy and sad, in equal measure all the way back to her formative years. Existing models largely accept that memories are prioritised and stored according to the emotional significance we give them, but AJ, and those like her, make us wonder if there's more to the system.

Proposing a Hypothetical Future Technology

Looking to the future, one would hope that diseases such as Alzheimer's and other forms of dementia would potentially be curable. Medicine and other forms of therapy could offer the answer, but neural implants may be a viable alternative.

As we have seen, different areas of the brain are responsible for different types of memories; some are conscious, some are not, some are unique to the individual, some are not. It is not possible to address them all at once, so specific examples must be selected.

Clinical Needs

There are two key questions to ask in developing a memory implant:

"What would we need an implant to do?"
"What can we realistically do?"

The former has virtually endless answers, but the latter has a much smaller cohort of answers. Pushing the boundaries of technology for the future is about taking answers to the first question and

seeing if we can make them answer the second. This is a good model for identifying areas for research, but not always for trying to develop a useable product here and now.

One of the unfortunate results of Alzheimer's and other forms of dementia is a loss of independence due to dangerous behaviour, such as leaving the front door open, leaving kitchen appliances on, etc[7,8]. These behaviours are one of the first reasons patients may need a caregiver, or to be moved into a home. If there was some way of mitigating such dangers, patients might remain living independently for that bit longer.

Concept

Berger et al.'s model took a specific behaviour and encouraged the rodent to perform a specific task with an electrical signal. In a similar manner, patients could be "taught" to avoid risky and dangerous behaviour such as leaving a lit gas ring unattended.

Having developed the neural activity as a MIMO signal, these signals could be programmed to a neural implant connected to a wireless receiver/transmitter; similar to cochlear implants, pacemakers, implantable cardioverter-defibrillators, deep-brain stimulation battery packs, etc.

The wireless device could also be controlled via specific signals, such as a proximity alarm. If the patient turns on a gas ring and leaves it unattended, a signal would be sent to the implant, and thus the brain, to go back and turn off the gas. Similarly, if a door was left open, and the patient left, a trigger for a MIMO signal to return and close the door would be generated. A wide range of signals to avoid danger could be programmed in this manner.

Looking to the future beyond these initial concepts, perhaps this could form the basis of a neurological reminder of important people in the patient's life. Not only is it distressing for family members to not be recognised by their own parents, siblings, etc., but one can only imagine the distress of the patient if they are constantly surrounded by people they perceive to be strangers.

It is likely, though not certain, that Berger et al.'s device signals a memory, not a forced set of actions, though it is unclear if the resulting actions of the rodents were completely autonomous. If the mechanism of Berger et al.'s system triggers a memory of a specific event as a prompt rather than a series of forced actions, this opens up a whole range of possibilities to "back up" other memories.

Benefits

While this would have no effect on the underlying disease process, it would have a significant impact both on the patient and their family/carers. It could provide peace of mind for all involved that, for now at least, the patient would be not likely to hurt themselves, and could be left unsupervised for significant periods of time. The possibility of more time to be trusted to be independent would likely also be very empowering and welcome amongst patients.

Long-term care of the dependent elderly, and concerns for their wellbeing, are well-documented sources of adverse consequences for carers. Depression, stress, poor sleep, anxiety, neglect of personal problems, health deterioration and other issues are commonly seen among the carer population[9,10].

Ultimately, most patients in this group end up in a nursing facility of some description, which is a heavy financial burden on the families, the state, or both. Generally, patients are happier at home, and if we can facilitate this for longer, everyone benefits both psychologically and financially.

Research Challenges

We are a long way from nearing anything resembling this technology being available for the general population. Prototypes of similar technology were first published in rodents only a year ago.

Research into the biology of memories needs much additional development.

Further *in vivo* studies and prototypes of the published neural implant, or similar, are needed, and then advancement to larger in vivo studies if successful. This is both costly and time-consuming, but given time, it is virtually inevitable.

Potential damage to neural tissue also needs to be considered. Potentially devastating side effects of surgical implantation or inadvertent stimulation of other areas is very possible, if not highly likely. This is a very important consideration when looking to apply this technology to humans.

Ethical Implications

Significant ethical hurdles would have to be overcome, particularly in the area of patient autonomy. Triggering a physical response/action in response to a wireless signal questions true autonomy. Whether or not the action is due to a memory prompt, or a direct neurological stimulation over which the patient has no control, is the big question. Neurological "reminders" rather than direct surrendering of bodily control are very different, and may not be possible to identify in rodent models.

Patient autonomy in a damaged neurological state, as prospective patients would be, is questionable. The validity of informed consent and appreciation of the risks in these patients are sometimes hard to determine, and in such an invasive procedure, is especially important to get right.

Due to the progressive nature of these conditions, this device would have an inherently short period of use. The time from diagnosis to required placement in a home or under full-time care could be only a few years. It begs the question of whether or not such a procedure is warranted.

However, if this technology were to progress to aid in recognition of family members and beyond as previously described, one could argue it never becomes obsolete.

However, one does need to bear in mind whether or not such an implant might fundamentally change the personality of the patient. For many people, their condition forms who they are, and major treatment in a non-immediately life-threatening situation may be somewhat unpalatable to many.

As with any wireless technology, the possibility of outside interference must also be considered. It has been found that pacemakers containing patient data can be accessed by an unauthorised operator, so privacy and protection issues would need to be safeguarded.

Ultimately, if we keep the patient's best interests at heart, and follow the four principles of autonomy, beneficence, non-maleficence and justice, we should find ourselves on the right path.

Conclusions

Memory is a complex neurological process, and science has barely scratched the surface. In light of recent discoveries and successful in vivo models, we can hypothesise on what possibilities lie ahead. No doubt there is a market, especially with a large proportion of the elderly population experiencing memory difficulties. In an ever-aging society, these issues are not going away. An implant of some description is inevitable, but how it might work, or when it may become a reality, we can only speculate.

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Preoperative Fasting: Closing the Gap Between Theory and Practice

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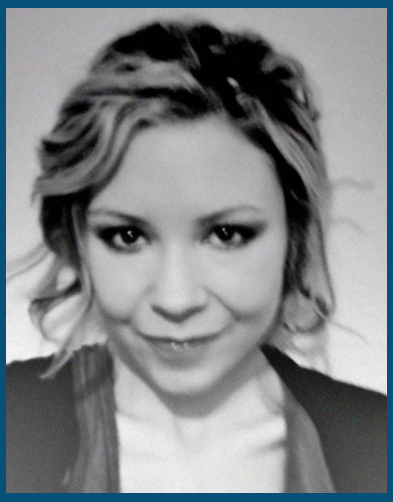

> "Patients should not be subjected to extensive fasting regimes. Despite this, traditional practices persist..."

Abstract

Prolonged preoperative fasting is experienced internationally, as healthcare professionals struggle to discard the traditional practice of "NPO" after midnight and implement new guidelines advocating shorter fasting times for patients. This literature review aims to address the continued practice of prolonged preoperative fasting and to provide a synopsis of the most up-to-date literature.

The researched literature included in-depth discussion of current guidelines and actual clinical practice, nursing and other healthcare professional perspectives, patient perspectives, and evidence-based practices. The author feels that further research examining the gap between preoperative fasting guidelines and actual practice would benefit the development of effective measures for change.

Introduction

Preoperative fasting requires all patients to fast absolutely from food and fluids for a specified minimum amount of time prior to anaesthesia. It is a necessary precaution to decrease the risk of pulmonary aspiration. It is an age-old tradition that began when Mendelson reported incidents of pulmonary aspiration during obstetric anaesthesia[1]. During anaesthesia, a patient's cough and gag reflex are affected, increasing the risk of pulmonary aspiration with potentially fatal consequences[2].

New evidence-based practices, which advocate shorter fasting times while ensuring no increase in patient risk, have been met with a reluctance by healthcare professionals to discard the trusted traditional practice[3]. The author developed an interest in the issue of prolonged preoperative fasting during work in various surgical specialities. It seems that, regardless of the speciality or the institution, prolonged fasting times can occur.

To examine the gap between theory and practice, a review of the literature was completed. Literature in the English language was compiled using the electronic databases CINAHL, PUBMED and COCHRANE, searching using the following key words: preprocedural fasting, preoperative fasting, patient anxiety and nursing use of evidence-based practice. Initially, it was hoped to limit the search to the past five years. However, it became evident that inclusion of guidelines from 1994 and literature published thereafter was necessary to ensure a comprehensive review. Inclusion of several older articles was deemed necessary to provide a historical overview of the topic.

The literature reviewed reflects the international multidisciplinary issue of prolonged preoperative fasting. Articles from across the globe were retrieved from nursing, anaesthetic and surgical journals, showing the prevalence of prolonged fasting and the determination to bring about change. From critically reading articles it became apparent that extensive fasting continues despite current research and guidelines. The findings will be discussed and critiqued under several headings: current guidelines and actual practices, nursing and other healthcare professional perspectives, patient perspectives and evidence-based practice.

Current guidelines and actual clinical practice

The literature revealed that the American Society of Anaesthesiologists (ASA) guidelines on preoperative fasting are the gold-standard reference for recommended safe fasting times[4]. Recommendations made for adults undergoing surgery who are otherwise considered healthy are as follows: patients should abstain from food for at least 6 hours and from clear fluids for at least 2 hours prior to surgery. Several national guidelines have been developed which draw on these guidelines[5,6].

Several quantitative studies have been carried out to determine whether shorter fasting times affect stomach volume and pH, thus posing a risk of pulmonary aspiration. One study investigated this by assigning participants to groups, each of which would receive 150 ml of fluid at different times preoperatively[8]. Stomach volume and pH were measured upon induction. Findings suggested that the gastric volumes and pH of all participants were comparable, regardless of the length of fasting. A similar study was conducted on patients who received unlimited amounts of clear fluid up to 3 hours preoperatively[9]. Further research was undertaken by the same author to determine if 300 ml of clear fluid preoperatively would affect the gastric volume and pH of obese patients[10]. Neither study found a significant difference between those who received fluids and those who did not.

A systematic review of the literature regarding preoperative fasting was conducted for the Cochrane database to establish the effects of various fasting regimes[11,12]. Review of 22 international trials revealed that there was no evidence to suggest that shortened fasting times would significantly alter the volume or pH of participants' gastric contents. In fact, it was observed that patients who received a drink of water up to two hours preoperatively had a significantly lower gastric volume than those who fasted from midnight. The stomach is never completely empty, even after an overnight fast, with a mean residual volume of 27 ml[13]. The Cochrane systematic review provides further evidence that traditional "NPO" from midnight is an out-dated, unfounded practice in need of modernisation.

In a 2002 study, participants were interviewed for 15 minutes by staff nurses to determine whether new ASA guidelines had changed preoperative fasting practices[14]. A semi-structured technique was used and patients' responses were compared to the ASA guidelines. The results showed average fasting times from fluids and solids of 12 and 14 hours, respectively. 97% fasted from fluids for over 6 hours, while extreme cases fasted from fluids for up to 20 hours. Disappointingly, 91% of patients claimed they received instructions to fast from midnight while only 28% reported receiving

information about the reasons for fasting pre-operatively. Several years later, data was compiled from two quality improvement (QI) data collection studies that had been conducted in 2000 and 2004 to monitor for improvements in practice. In both studies, patients were interviewed after surgery[15]. Several patients could not take part in the interviews as they complained of drowsiness, which may call into question the accuracy of the findings collected from the remaining patients. They found that from 2000 to 2004, patients' instructed fasting times from fluids had decreased by 0.11% while actual fasting from fluids saw a drop of 0.005%, from an average of 11.9 hours to 11 hours. The instructed fasting times for solids had improved from an average of 10 hours in 2000 to 9.7 hours in 2004. Actual fasting times from solids only decreased marginally from a mean of 14.5 hours to 14.2 hours in 2004. Regardless of the small changes noted, this research has shown that although progress may be slow, it is happening and can continue with the hard work of healthcare professionals using a multidisciplinary approach.

Nursing and other healthcare professional perspectives

Qualitative research regarding nurses' knowledge and perceptions of preoperative fasting identified that a lack of knowledge, ritualistic practice and unsmooth running of intra-unitary systems resulted in prolonged patient fasting[16]. All participants understood the importance of fasting in preventing vomiting and aspiration perioperatively. Surprisingly, when questioned about appropriate fasting times, only two of fifteen nurses expressed knowledge of the ASA guidelines while the majority overestimated the required fasting durations by two hours. Upon further questioning it was discovered that recommended fasting times as set out by the ASA are rarely adhered to and that patients on the afternoon list often endured "NPO" status for up to 12 hours.

A similar phenomenological project in America used open-ended questions aimed at interpreting nurses', patients', and anaesthetists' perceptions of preoperative fasting[17]. The rationale governing the need to fast was known by the majority of those surveyed: 73% of nurses correctly identified the risk of vomiting and aspiration as the main safety concern. However, healthcare professionals admitted that leaving patients fasting from midnight was common practice as part of an effort to avoid patient non-compliance and surgical rescheduling issues. Interestingly, only 13% of nurses could correctly refer to the guidelines set out by the ASA in 1999. Recommendations for change include staff education and changes in hospital policy regarding acceptable fasting times.

Patient perspective

Nurses spend the most time with patients and are ideally placed to observe both the physical and psychosocial effects of preoperative fasting, including irritability, confusion and the social isolation of missing meal times[3]. Prolonged fasting increases the risk of developing hypoglycaemia, dehydration, electrolyte imbalances, headache and confusion[18]. In patients with poor preoperative nutrition, recovery may be complicated by poor wound healing, increased risk of pressure sores and increased likelihood of experiencing nausea and vomiting postoperatively when fasted excessively[19, 20].

Nurses and anaesthetists believe that patients lack sufficient knowledge regarding the rationale for preoperative fasting, resulting in confusion about the fasting lengths and contributing to issues of non-compliance[17]. Further evidence supporting the existence of this belief comes from a qualitative study, which found that the belief that patients will not understand or comply with fasting instructions often leads to longer fasting times[21]. Anaesthetists and surgeons feared that patients might get confused and consume solid food if they were told they could ingest clear fluid for longer[21].

A more comprehensive quantitative research study focusing exclusively on patients' knowledge and understanding of preoperative fasting required 100 elective day surgery patients to complete a short

questionnaire before discharge[22]. 63% of patients reported having been seen by the anaesthetist at the pre-assessment clinic and the remaining were seen before surgery. In addition, the majority of participants reported receiving information from nursing staff and 90% had received written information. Despite this, the average fasting times were greater than those recommended in the ASA guidelines, with an average fasting time from solids of 10.8 hours and 6.5 hours from fluids. In an attempt to explain these prolonged fasting times, a question was included specifically to access participants' comprehension of the reasons for fasting before surgery. Disappointingly, only 22% provided a correct response. This may have been due to the fact that, for a response to be considered correct, participants had to allude specifically to the risk of pulmonary aspiration. Encouragingly, actual compliance was high and 65% agreed that following fasting instructions was important. The results of this study suggest that improvements in patient education are necessary. It is reasonable to conclude that more research needs to be compiled about how healthcare professionals can improve patient education, conveying the importance of adhering to fasting instructions without causing unfounded fear and anxiety about the risk involved.

Evidence-based practice

An Bord Altranais demands that nurses provide the highest standard of patient care by overseeing the implementation of evidence-based fasting practices[23]. The evidence provided suggests it is advisable that patients should not be subjected to extensive fasting regimes. Despite this, traditional practices persist due to preconceived ideas about patient knowledge, changes in theatre lists, and lack of knowledge about up-to-date research and guidelines. Nurses are at the forefront of patient care and have a responsibility to ensure best practices are followed. Many nurses feel that their awareness of current research is inadequate, blaming a lack of physical access to the research[16]. Nurses can only advocate for the patients' best interests when they are aware of current best practices and recommendations[24].

Action research is a practical and user-friendly research method for nurses[25]. Identification of excessive preoperative fasting as a problem and isolation of probable contributing factors, including lack of knowledge and changes to theatre lists, was the first stage in an action research project aimed at making improvements in preoperative fasting procedures in a UK hospital[3]. A primary quantitative study was designed to test the extent of the problem in the chosen orthopaedic ward. A convenience sample of 110 patients was included in the study, which required the patients' nurse to complete a questionnaire gathering data about average fasting times endured. The study confirmed that prolonged preoperative fasting was occurring, triggering the development of a double-approach action plan. Anaesthetists were required to prescribe the shortest possible fasting times while the researchers implemented an education programme on preoperative fasting to nursing staff. A questionnaire at the end of these sessions assessed the level of nursing knowledge. Posters and information packs were placed on the ward for easy reference. On completion of the educational programme and implementation of the changes by the anaesthetists, a repeat of the initial questionnaire, with minor additions, was completed using another convenience sample of 106 orthopaedic patients to determine whether fasting times had been reduced. The results showed that mean fasting time was reduced by 5.4 hours, bringing the average patient fasting times down to 6.54 hours, more in line with the new ASA guidelines. Action research, as proven by the success of this action project, is an accredited method of implementing health innovations[26]. This action project was successful in facilitating improvements in preoperative fasting practices, developing nursing knowledge, and encouraging enhanced communication between disciplines.

The findings of two separate hospital-based audits revealed that patients were subjected to prolonged preoperative fasting[27,28]. The first, conducted on a sample of 140 orthopaedic patients, revealed the possibility of changes in the order of the operating

list as the main barrier faced. A trial of 10 patients was devised in response, allowing all patients to drink at 6am and then allocating individualised fasting regimes after all preoperative checks were complete[27]. Personalised fasting times were estimated by the nurse in charge of the operating list and communicated to the ward nurses. Patients were allowed fluids up to two hours before the estimated surgical slot. However, the duration of some operations was underestimated, resulting in seven of the ten patients fasting from fluids for an average 3.5 hours. The second audit focused on the impact of improved communication between staff and patients regarding fasting times. The results showed a decrease in fasting times after improvements in communication and education[28]. The results of these trials prove that through continued auditing, change in preoperative fasting practices can occur.

Conclusion

The literature examined demonstrates the relevance of preoperative fasting as a topic for discussion. In order to fulfill An Bord Altranais' goal of implementing the most current evidence-based nursing practices in Irish hospitals, the nurse needs to discover a mechanism of introducing the preoperative fasting recommendations made by the ASA. By ensuring a thorough historical and scientific understanding of the rationale behind preoperative fasting, the nurse can prevent patient harm by decreasing anxiety, discomfort, hunger, and thirst.

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What the PHECC: An Introduction to Pre-Hospital Emergency Care

Jack Collins, 4th Year Medicine, UCC

> "The current evidence base suggests that doctors have a beneficial and worthwhile role in pre-hospital care."

Abstract

Surprisingly, the way in which pre-hospital emergency care is delivered varies greatly internationally. Two contrasting models can be identified: one physician-led, the other with direct or indirect medical supervision. The development of pre-hospital care in Ireland has been convoluted and stunted. However, in less than a decade, the Irish emergency medical services have evolved significantly. Currently, three clinical levels of pre-hospital care practitioner exist in Ireland: emergency medical technician (EMT), paramedic and advanced paramedic. Advanced paramedics are the only level considered to be advanced life support providers. The levels were created following the establishment of a national emergency medical services regulator known as the Pre-Hospital Emergency Care Council (PHECC). PHECC aims to protect the public by developing education standards, maintaining the register of pre-hospital care practitioners and enforcing a code of professional conduct. PHECC have also introduced clinical practice guidelines, which direct how acutely ill or injured patients should be treated by pre-hospital care practitioners. In many jurisdictions, doctors play a pivotal role in the delivery of pre-hospital care, a specialty known as "immediate care" in some countries. There are few examples of doctors in Ireland providing formal pre-hospital care, despite evidence suggesting that the involvement of medical practitioners in the pre-hospital setting is beneficial even in the presence of other advanced life support providers.

Introduction

Pre-hospital care is defined as the provision of skilled health care at the site of a traumatic incident or medical emergency and encompasses those competencies delivered by appropriately trained clinicians[1]. The administration of emergency medical care outside of hospitals dates back to the late 18th century and has military origins[2]. Dominique Jean Larrey, Napoleon's chief surgeon

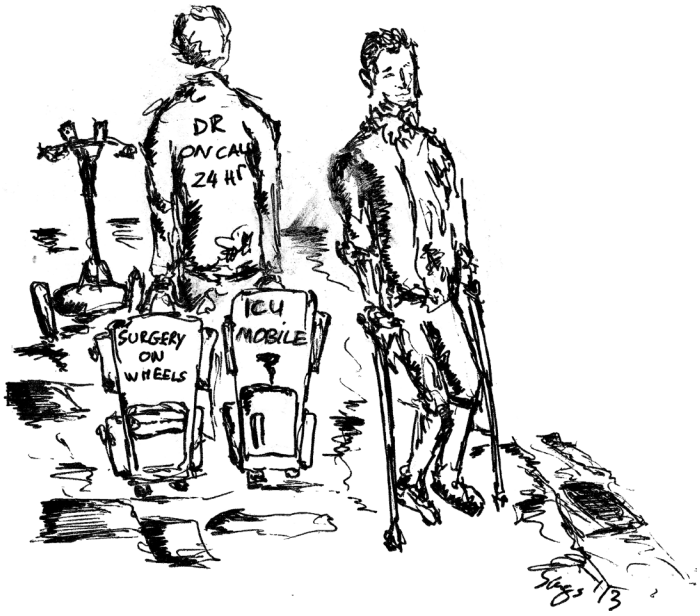
and a pioneer in battlefield medicine, established the first horse-drawn 'flying ambulances' to care for wounded soldiers. Care of injured soldiers was revolutionised in this period, as it saw the introduction of the first military division with responsibility for medical care. Larrey also introduced the concept of triage to the battlefield, which ensured that patients received care according to need. Since the French revolution, pre-hospital care has evolved significantly and is now considered a subspecialty of emergency medicine in many jurisdictions[3-5].

Models of Pre-Hospital Care

The manner in which acutely ill or injured patients are cared for in the pre-hospital environment varies greatly from country to country. Internationally, there are two recognised pre-hospital emergency care models, namely the Franco-German model and the Anglo-American model, which differ significantly in many respects.

Table 1 is adapted from Al-Shaqsi[6] and demonstrates the main differences between the models.

A notable example of the Franco-German model is the French Service d'Aide Médicale Urgente (SAMU), which came into existence in the 1960s, providing some of the world's first formal physician-led pre-hospital care. In this system, all emergency calls seeking medical care are directed to a SAMU call center where a medical practitioner performs



Broken leg? If only it were a challenge for Dr Segway

a detailed assessment and triage leading to four possible outcomes:

1. Advice and discharge
2. Advice to attend a hospital or clinic
3. Dispatch of a general practitioner or SAMU-affiliated physician team
4. Dispatch of a mobile intensive care unit (ambulance or helicopter)

In the SAMU system, the primary pre-hospital care provider is always a medical practitioner who can provide various advanced critical-care interventions such as rapid-sequence induction, intraosseous cannulation and needle cricothyroidotomy. Ancillary staff do not engage in any form of advanced life support (ALS) and the role of paramedic, as we know it, does not exist. This model of pre-hospital

Table 1. Comparing Anglo-American and Franco-German models of pre-hospital emergency care

Anglo-American	Franco-German
Few treated and discharged on scene. Majority transported to hospital.	Many treated and discharged on scene. Referral and follow-up where appropriate. Avoids unnecessary transport to hospital.
Paramedics operating under the indirect supervision of medical practitioners or following clinical practice guidelines.	Medical practitioners supported by paramedics, registered nurses or ancillary staff.
"Scoop and Run"	"Stay and Play"
Bring the patient to hospital.	Bring the hospital to the patient.
Emergency Department	Direct to appropriate ward e.g. Cardiac Care Unit, bypassing Emergency Department

Al-Shaqsi[6]

care claims to provide both rapid and effective medical care, promote patient comfort and ensure the best utility of resources[7]. A recent paper discussing the provision of pre-hospital care in Shanghai reports that, similarly, all ambulances are staffed with an emergency physician[8].

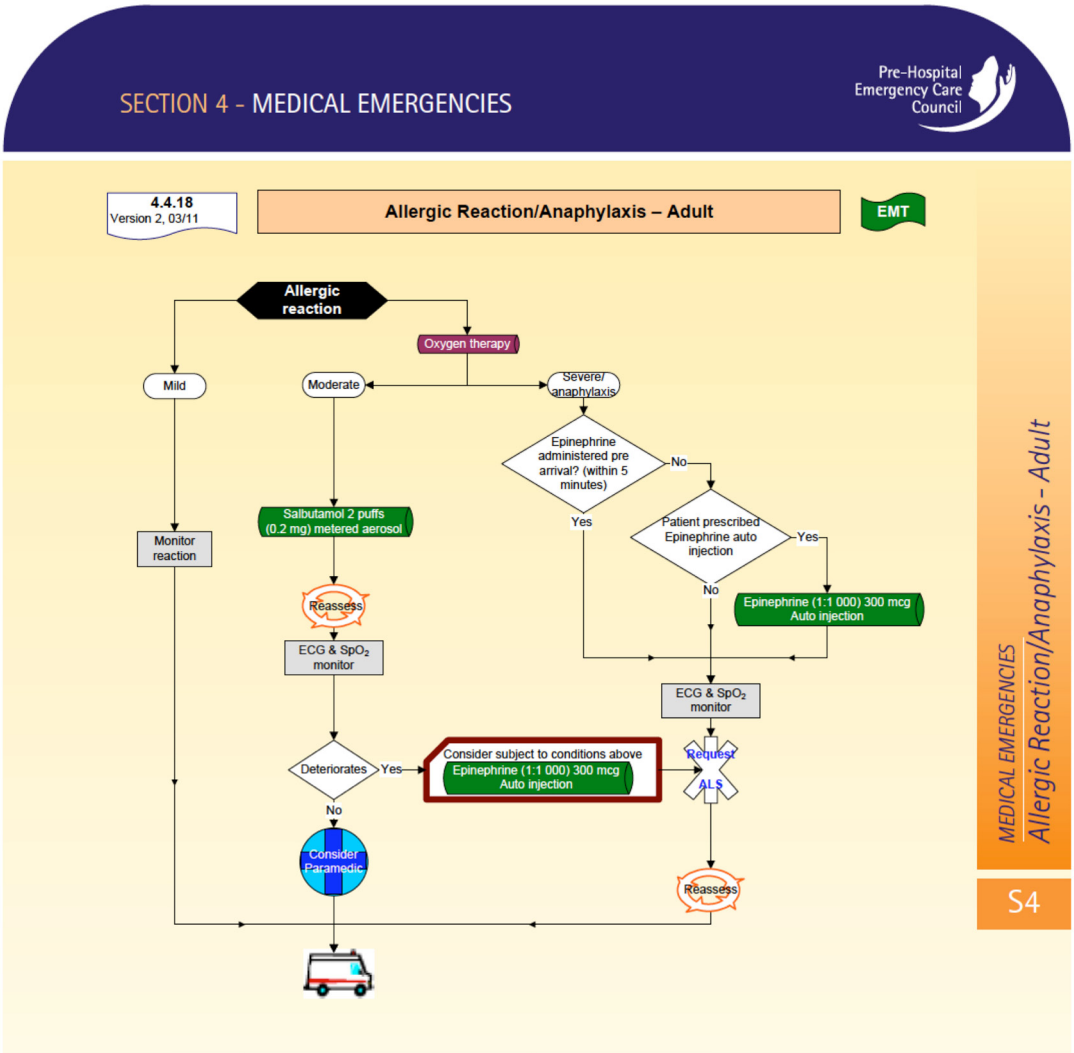


Figure 1. Sample Clinical Practice Guideline. Pre-Hospital Emergency Care Council, Clinical practice guidelines: emergency medical technician: 2012 edition, Ireland, (2012).

Conversely, pre-hospital care in the United Kingdom (UK) is provided by pre-hospital care practitioners, which is in keeping with the Anglo-American model. An emergency call to the ambulance service is dealt with by an ambulance controller and results in the dispatch of an ambulance with a paramedic and ambulance technician crew. A recent UK study indicated that, with the exception of London city, physicians are not employed by the National Health Service for the purpose of pre-hospital care[9]. However, many medical practitioners are involved in providing pre-hospital care through charitable organisations such as the British Association for Immediate Care (BASICS). The majority of their work involves responding to motor vehicle collisions or other serious accidents as immediate care doctors. Medical practitioners formed part of the crew in ten out of the fourteen helicopter emergency medical services (HEMS) that existed in the UK in 2007[10]. However, in line with the Anglo-American model, the UK has moved towards an extended role for paramedics with the introduction of emergency care practitioners. Known as paramedic practitioners, they operate as autonomous professionals, accountable for their clinical practice. They can perform advanced procedures such as suturing of wounds, and may prescribe medications such as antibiotics, a privilege normally reserved for medical practitioners[11].

What the PHECC

Historically, the provision of pre-hospital care in Ireland was disorganised and somewhat haphazard. Ambulances were dispatched from hospitals with a porter and nurse team, providing rudimentary pre-hospital care. In 1997, the single clinical level of EMT was introduced. This was the first step towards formalised training for ambulance personnel. At this time, general practitioners were often summoned to the scene of an accident, or perhaps called to

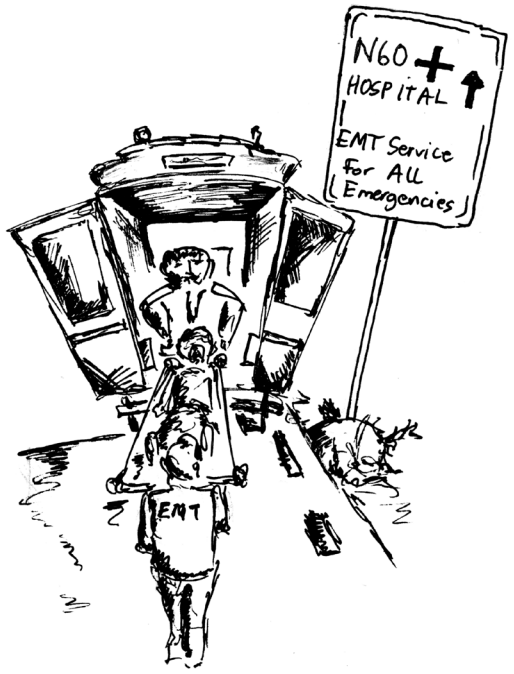
Skill/Clinical Procedure	EMT	P	AP
Airway & Breathing Management			
BVM	✓	✓	✓
Cricoid pressure	✓	✓	✓
FBAO management	✓	✓	✓
Head tilt chin lift	✓	✓	✓
Jaw thrust	✓	✓	✓
Non-rebreather mask	✓	✓	✓
OPA	✓	✓	✓
Oxygen humidification	✓	✓	✓
Pocket mask	✓	✓	✓
Recovery position	✓	✓	✓
SpO_2 monitoring	✓	✓	✓
Suctioning	✓	✓	✓
Venturi mask	✓	✓	✓
Flow restricted oxygen-powered ventilation device		✓	✓
LMA/LT adult		✓	✓
NPA		✓	✓
Peak flow		✓	✓
End Tidal CO_2 monitoring			✓
Endotracheal intubation			✓
Laryngoscopy and Magill forceps			✓
LMA/LT child			✓
Nasogastric tube			✓
Needle cricothyrotomy			✓
Needle thoracocentesis			✓

Figure 2. *PHECC skill matrix. Matrix indicating the procedures which each level of pre-hospital care practitioner is authorised by PHECC to perform. Pre-Hospital Emergency Care Council, Statutory registration and pre-hospital emergency care practitioners, Ireland, (2009).*

the home of a patient, but no formal arrangements existed. The establishment of the Pre-Hospital Emergency Care Council (PHECC) in 2000 resulted in the introduction of education and training standards, clinical audit and a code of professional conduct in the Irish pre-hospital environment[12].

At present in Ireland, pre-hospital care is commonly delivered by a group of pre-hospital care practitioners regulated by PHECC. Their register is subdivided into three clinical levels: EMT, paramedic and advanced paramedic. These healthcare professionals are authorised to perform interventions and administer medications according to Clinical Practice Guidelines (CPGs) and function without the direct supervision of a medical practitioner (see Figure 1, sample CPG). They may perform many interventions for acutely unwell or injured patients, depending on their training and level on the PHECC register (see Figure 2 for PHECC skill matrix). Advanced paramedics are the only level authorised to offer ALS to patients. Most pre-hospital care practitioners provide care through their employment with the statutory ambulance services administered by the Health Service Executive (HSE) or the Dublin Fire Brigade. However, some practitioners have trained and operate solely within auxiliary or voluntary organisations such as the Civil Defence, St John Ambulance or the Order of Malta. Similarly, registered nurses, operating within their scope of practice, may deliver care in the pre-hospital environment and often do so with such organisations.

Currently, there is no formal national arrangement for the regular involvement of medical practitioners in pre-hospital emergency care in Ireland. Surgical teams may be summoned to accidents when emergency surgical interventions are necessary, but this is on an ad hoc basis. The Emergency Department (ED) at Cork University Hospital (CUH) piloted a pre-hospital physician response team in conjunction with the HSE ambulance service and found pre-hospital physicians to be beneficial despite the recent deployment of advanced paramedics[13]. There are some sporadic examples of local arrangements between doctors with a



Right, we can't fix this papercut. We're on the way to A&E!

Clinical Points

1. Pre-hospital care is considered a subspecialty of emergency medicine in many jurisdictions.

2. The delivery of pre-hospital care in Ireland is led by a group of registered healthcare practitioners known collectively as pre-hospital care practitioners.

3. The Pre-Hospital Emergency Care Council regulates the three levels of pre-hospital care practitioner: emergency medical technician, paramedic and advanced paramedic.

4. There is a paucity in the involvement of medical practitioners in pre-hospital care in Ireland, in stark contrast to many other developed countries.

5. A limited amount of current literature suggests that the involvement of doctors in the management of patients in the pre-hospital environment is beneficial.

special interest in pre-hospital care in Ireland, and the author encountered much anecdotal evidence of their successful involvement in cases providing advanced interventions such as surgical airways and rapid sequence induction, described by one as "bringing the ICU to the roadside". Unfortunately, there is no Irish evidence to suggest whether the addition of medical practitioners to the national ambulance service would improve morbidity or mortality following acute illness or injury. The national ambulance service is currently piloting a national aeromedical response team where a single helicopter is staffed by one advanced paramedic and one EMT. Perhaps the addition of an emergency physician to this crew could further improve the treatment of patients requiring immediate medical care.

Are doctors needed?

In 1986, Pepe, a pioneer of emergency medicine and pre-hospital care, described the work of paramedics as "the practice of medicine through physician surrogates"[14]. Despite the introduction of advanced life support providers, such as advanced paramedics, recent data indicates that the involvement of physicians in pre-hospital care has a significant impact on patient morbidity and mortality.

One systematic review of 26 studies found that physician involvement in pre-hospital treatment correlated with increased survival from traumatic injury and myocardial infarction when compared to paramedic care alone[15]. Similarly, an Australian study found that the addition of a medical practitioner to a HEMS team significantly decreased the mortality of blunt trauma patients when compared to paramedic care. It was also discovered that the critical "on-scene" time was not increased by physician presence[16]. This data is supported by a study from the UK, which found that 'on-scene' time was not prolonged by physicians despite advanced medical interventions being provided[17]. A Cochrane systematic review conducted in 2010 concluded that the provision of ALS in the pre-hospital environment by non-physicians offered no benefit to patients[18], while a more recent British paper identified that doctors have a higher rate of successful intubation on the first attempt when compared to critical care paramedics[19]. It is clear that a paucity remains in data comparing the performance of pre-hospital physicians to non-physicians; however, the current evidence base suggests that doctors have a beneficial and worthwhile role in pre-hospital care.

While it is unlikely that physicians will be extensively deployed within the Irish ambulance service, the development of immediate care schemes similar to BASICS is a reasonable expectation. Already, Munster has seen doctors engaged with West Cork Rapid Response attending motor vehicle collisions on isolated, rural roads. Recently the UK College of Emergency Medicine began to recognise pre-hospital care as a

subspecialty of emergency medicine. This indicates a desire to further develop pre-hospital physician-delivered care and implement formal training in this field, even in a country whose statutory pre-hospital care model is much less grounded in physician-led care.

Conclusion

Physician-delivered pre-hospital care remains an underdeveloped resource in Ireland. Much debate continues as to the benefit of immediate care doctors; however, most commentators conclude that the provision of advanced life support by non-physicians is enhanced by the support and presence of a medical practitioner. It is clear that pre-hospital care practitioners have a defined skill set, which can be complemented by physician presence. Research is required to determine the need for pre-hospital physicians in Ireland, as advanced paramedics already provide a high level of care. However, the performance of these practitioners is yet to be assessed and current evidence suggests that physician presence remains beneficial even when other ALS providers are operational.

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At a Time of Change

Winning Essay, Sheppard Memorial Prize, 2012

Deirdre Kelly, 4th Year Medicine, TCD

> **"General practitioners are on the "front line" of medicine. They are the patient's advocate in the health system."**

"In its most highly developed form, primary care is the point of entry into the health services system and the locus of responsibility for organising care for patients and populations over time. There is a universally held belief that the substance of primary care is essentially simple. Nothing could be further from the truth."[1]

This quote is from the recently deceased Barbara Starfield, Professor of Health Policy and Management at the Johns Hopkins Bloomberg School of Public Health, who advocated for excellence in primary care programmes both in America and all over the world.

In an ideal world, the health system would be structured in the way Barbara Starfield suggests. The general practitioner is the "gatekeeper", the first port of call, preventing unnecessary hospital visits and providing continuous and personal health care. In an ideal world, the general practitioner is a separate entity to any administrative or government body and advocates for patient care alone. In an ideal world, the general practitioner charges an appropriate fee for the private consultation and in an ideal world, everyone can afford this bill. Unfortunately, we do not live in an ideal world.

From a purely economic standpoint, however, general practice has shown its virtue. The higher the percentage of primary care physicians, the lower the cost of health care in a country. 70% of UK doctors work in primary care and only 6% of the British gross domestic product (GDP) is spent on healthcare. This compares to only 30% of doctors in the USA, Barbara Starfield's native land, employed in primary care and the resultant 12% of the GDP allocated to healthcare[2]. The more general practitioners there are providing a community service and an alternative to the hospital for many conditions, the better the chance of keeping healthcare expenditure in our country lower.

This, however, says very little about the inefficiencies and inadequacies of the Irish healthcare system. Far from this ideal world, there are those in the community who cannot afford to visit their doctor: those who fall just short of the requirements needed for a medical card. Severely disadvantaged areas are short of general practitioners and those patients who cannot afford to, or otherwise cannot access a GP, are the same people at a higher risk of developing chronic disease and multiple morbidity. They are the same people who will inevitably show up in A&E again and again, presenting at a later stage of disease and therefore at a more difficult stage to treat, diminishing their chance of a good prognosis and, in the most cynical way of thinking, costing society more to treat them.

In 1971, Dr Julian Tudor Hart coined the phrase "The Inverse Care Law"[3]. Interpreted simply, this states that those in most need of health care in society have the least access to it. It is a well-known fact that the less well off individuals in the community are also those with poorer health due to many contributing factors. These include income inequality, lack of government spending, lack of social supports, lifestyle factors such as smoking, poor diet and lack of exercise and other non-medical factors such as housing and transport. Those in deprived areas are less likely to complete their secondary education or progress to third level, limiting their career opportunities and income. Many are left unemployed and those who have work often have poorer training and working conditions which in themselves are health risks.

Drug abuse is more common, as is alcoholism, and the social support is not in place to encourage change or even prevention of these serious issues. Along with these factors comes poor nutrition. As healthy food is becoming more expensive and processed food comparably cheaper, obesity is rising in the poorer communities in Ireland. An in-depth report by Combat Poverty Agency in 2008[4] revealed that due to these factors, "almost half (47%) of those who were consistently poor (ie. in income poverty and experiencing deprivation) and 38% of those who were income-poor reported having a chronic illness, compared with 23% of the general population".

Not only did they describe the health effects of poverty on those of low income, they also outlined the health issues facing the marginalised groups in society, including the travelling community, asylum seekers, those suffering from mental health issues, those of a different sexual orientation, those with disabilities and the homeless. These people, while affected by many of the issues facing those of a low income in a disadvantaged area, also have distinct health issues of their own. For example, asylum seekers may not have had the privilege of vaccinations that we are so lucky to receive and may be more at risk of infectious diseases, whereas homeless people have a higher incidence of tuberculosis and those with Down syndrome commonly suffer with cardiac issues. Many in these groups will also by stigmatised due to prejudices in society, such as those who suffer with schizophrenia or those in the travelling community.

General practitioners are on the "front line" of medicine. They are the patient's advocate in the health system. If an issue can be dealt with in the practice, the doctor can put worries to rest, instilling trust and furthering the doctor-patient bond. Thus, if an issue needs a more specialised opinion and the patient must delve deeper into the health system, they will know that their GP is their navigator and with them 100% of the way, charting their journey and hopefully their recovery. Luckily in Ireland, I believe we have such GPs. I have seen first-hand, while on placement in primary care, GPs taking the necessary time, listening closely to what really concerns the patient and then speaking on their behalf, all the while with an interest in the patient's well-being above all else.

Currently, the Irish government is bringing in changes to improve healthcare as part of the Programme for Government 2011. With regard to the plans for primary care alone, plenty of changes

are sought. Universal free health care is to be implemented. GP fees will be eradicated for all. GPs will have greater access to diagnostic equipment to alleviate pressure on the hospitals. The number of places on GP training schemes will be increased to allow greater numbers of GPs in practice. Similar measures will be implemented for primary care nurses and other professionals in the sector, including psychologists and counsellors, to aid in the management of mental health in the community.

There will be four phases to this plan. First, those with long-term illnesses will be granted free health care. Then, claimants of free drugs under the High-Tech Drugs Scheme will receive free health care. Thirdly, health care for everyone else will be subsidised and finally, primary care will be free for all. Under this plan, GPs will be expected to work in a multidisciplinary primary care team. GPs will then be financed entirely by the government[5].

Doctors from areas of deprivation seem to welcome these changes. Dr Edel McGinnity, whose practice is in Mulhuddart, Co. Dublin, stressed how much help being part of a primary care team has helped her practice and how her patients would benefit from a greater access to healthcare[6]. Advocacy for her patients is her simple underlying conviction. She also stated how important it is for general practitioners to advocate for themselves. The Irish College of General Practitioners has welcomed many of the recommendations made by former junior minister for primary care, Roisin Shortall, such as ring-fencing a primary care budget and access to diagnostics for GPs[9]. Suggestions have been made in relation to the restructuring of general practice into primary care teams, a change welcomed by Dr McGinnity and other GPs in areas of deprivation. The ICGP report suggests, however, that "engagement at local level planning and developing services needs to be undertaken and it must be acknowledged that one size does not fit all"[7].

This perhaps applies to many primary care physicians who do not practise in disadvantaged areas, who may have invested a huge sum of money into setting up efficient and well-run practices and who have developed the same trusting, personal relationships with their patients, who have connected with other healthcare professionals in an informal way, finding this as effective as the proposed structured primary care teams. These GPs have systems in place for long-term care management and prefer to be autonomous. They worry ablout being entirely under the authority of the HSE. This, I believe, is completely reasonable and healthy. The Programme for Government is aimed at helping those less well off in society and this is very desirable. However, some GPs fear these developments will actually diminish the level of health care currently available. Some GPs feel that attending public care team meetings will reduce patient contact time by 1.5-2 hours per week and that this is an inefficient use of GP time and resources, increasing inpatient admission rate by 2.5%, outpatient visits by 2%, emergency department attendances by 4% and surgeries by 3%, thus costing more and putting more pressure on the hospital services, reversing the desired effect of this reform[8].

There are fears among GPs of how disenfranchised they could potentially become with the HSE (or whatever new body would be set up under the Programme for Government) taking over the regulation of primary care, which has traditionally remained quite independent of government management. This will severely limit the GP's role as an advocate for their patients. A worrying element of the planning of these changes is that the implementation group involved in coordinating the logistics of universal healthcare is made up of those with experience in public service administration or academics but only one doctor, a pathologist, has had an input[8]. It is understandable for many GPs to be anxious about a major upheaval of the current system, which does work effectively in many areas, without any consultation from those with a background in general practice. It is especially unsettling to see rifts within the Department of Health that have led to Ms. Shortall's resignation. With such an extensive remodelling of primary care

in prospect, it would be far more reassuring to see a united team working together for the good of the patients and the GPs rather than internal strife and controversy. Much of the focus seems to be on the policy-makers rather than those who will be affected by the end result.

These fears are supplemented by reports from the IMO that these new developments are not in the patients', or indeed in the GPs', best interests. Recent reports that the primary care centres planned to be opening in the south of the country show no signs of new diagnostic developments, no improvements of the current primary care facilities, no signs of a shift of hospital services to the community and that the doctors who enter into the new system would have no tenancy rights and be in constant danger of the HSE dismissing their services[9]. There seems little sense in reshuffling GPs from perfectly good practices they have set up to properties rented by the HSE if there are no advantages to patient care, especially if their livelihoods become more precarious and they are under the authority of a body which has no GP representation. Additionally, if the government is the only financier then the system becomes a cartel and GPs lose their independence and advocating role. What the government also has to be careful with is the future recruitment of newly qualified primary care workers. In recent times, we have seen a dramatic increase of junior hospital doctors leaving Ireland for better working conditions and pay abroad at the expense of hospital services here. The work of general practice should attract the brightest and best of the qualifying doctors to continue to improve primary care for the citizens of Ireland.

In order to do this, the first step the government needs to take is to involve the general practitioners to a greater extent and listen to their opinions on reform and universal healthcare. As worthwhile and commendable project as it is, it cannot be blanketed all at once across a country with such varying socio-economics. Any developments made need to be made at a local level, rather than a national level, in places that need and welcome change. Universal free health care is a wonderful idea but the implementation of primary care teams and actual improvements in services require more co-ordination between those proposing the change and those on whose livelihoods it would impact, who continually provide excellent primary care facilities and services to their patients. General practice in Ireland has extremely short waiting lists, there is no two-tier approach to public and private patients and they resist medical inflation, so rather than restructuring primary care the government should perhaps build on the good, albeit not perfect, system which already exists.

In a country as developed and progressive as ours, it is deplorable that there are some who will not seek medical advice for financial reasons. For the good of any individual and for the country as a whole, this is a situation that badly needs to be rectified and few can argue against free healthcare. It is a commendable step that our country is taking to eradicate this problem. However, the Programme for Government is such a broad and ambitious strategy that I am afraid this worthy undertaking will be lost in favour of a different, more easily implemented task. I believe primary care teams are, in theory, a wonderful way for the various aspects of health care to interact and help each other, and in many situations would be extremely welcome. But for this to be the first step in our healthcare reform, before universal health care has been brought in and without primary care input, seems like poor prioritising. It is oversimplifying the situation to reroute every GP into primary care teams when many doctors work out of perfectly functional centres with unforced arrangements with their local physiotherapists, counsellors etc. already in place. Perhaps it is time for the government to listen to those at the front line, who know how primary care should work and what is needed at a local level, and then perhaps money can be spent where it is needed and to do away with the inverse care law in Ireland.

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“An overemphasis on patient autonomy results in patients feeling abandoned and physicians feeling frustrated.”

Winning Essay, Jeremy Swan Medal, 2012

Iain McGurgan, 4th Year Medicine, TCD

“In the steady shift of medical practice from its paternalistic roots to patient-centrality, respect for patient autonomy has come to overshadow its fellow principles of beneficence, non-maleficence and justice.”

“What would you do, doctor?” an 84-year-old lady asks the consultant, for the third time, in the outpatient clinic on a November morning of last year. She is faced with the choice of undergoing surgery for her worsening cataracts, which may also relieve some intraocular pressure to address her early glaucoma, or to proceed with medical management for the glaucoma alone. On receiving an identical measured response she turns to me, the medical student in the corner, as a last resort for decisive intervention. I avoid her gaze, and avoid the mounting conflict: a conflict not between a doctor and a patient, nor between their respective interests, but between two of the great ethical cornerstones of medical practice: autonomy and beneficence. As if to illustrate for the consultant and me the two extremes of moral standing on the much-debated matter of respect for autonomy, the very next patient to enter the consultation room is a 65-year-old man with diabetic retinopathy and an unfortunate host of co-morbidities. He thrusts his hat on to the table and proclaims, before even a word of greeting, “Let me tell you now, you won’t play God with me!”

The word autonomy derives from the Greek words for self-governance. It encompasses a capacity to decide and act without the constraints of controlling interferences by others or personal limitations, most notably a lack of adequate understanding, which prevent meaningful choice[1]. In the steady shift of medical practice from its paternalistic roots to patient-centrality, respect for patient autonomy has come to overshadow its fellow principles of beneficence, non-maleficence and justice. To quote the American bioethicist, Paul Wolpe: “for better or for worse, autonomy has emerged as the most powerful principle in bioethics, the basis of much theory and much regulation, and has become the ‘default’ principle”[2]. But what has triggered the emergence, and arguably the overemphasis, of autonomy as the primary governing principle in

medical ethics? Wolpe has suggested that as a result of the "erosion of trust" between the doctor and patient (a reflection of the documented widespread decline in social trust and trust in the medical profession since the 1960s), rituals of trust in the form of dialogues clearly establishing the patient's autonomy have emerged as a substitute for organic trust[3]. Furthermore, respect for patient autonomy may actually reinforce physician authority rather than impede professional privilege, as, in reality, autonomy tends to be limited to a right to refuse a particular treatment rather than to demand it. Others argue that by prioritizing patient autonomy, the doctor shifts the burden of decision-making to the patient and thus is relieved of some responsibility, as well as being less likely to be sued for malpractice[4].

Regardless of the reasons, the unprecedented prominence ascribed to the respect for autonomy has led to momentous alterations in the dynamic of the doctor-patient relationship. The paternalistic model sees the doctor as being in a better position than the patient to decide what is in the patient's best interests, and thus allows the doctor to act accordingly even if this contradicts the expressed wishes of the patient[5]. Other models presented by Emanuel and Emanuel[6] involve respect for autonomy in varying degrees, ranging from the informative model, where the doctor acts only to provide medical facts, to the interpretive and distributive models, wherein discussion about management is encouraged and patient values can be challenged to an extent (in the latter)[6]. It is clear that patient autonomy only can be completely respected in the informative model. Thus, I believe it is the move towards incorporating this model in clinical practice with competent patients, to maximally distance ourselves from paternalism, which has left these patients feeling unsupported. It seems, in the case of patients like the 84-year-old lady above, that it is the unfulfilled want for reinstatement of the primacy of beneficence in the doctor-patient relationship that results in the frustration and abandonment experienced by doctors and patients, respectively. But surely accommodating this would send us straight back to the dark era of unopposed paternalism? Not if we adopt an alternative model, as presented by Edmund Pellegrino (an avid defender of the prominence of beneficence), of autonomy incorporating rather than replacing beneficence. He argues that "the best interests of the patients are intimately linked with their preferences"[1]; the patient's wishes alone determine the extent of the doctor's beneficent role, even if this wish involves a rational request for the doctor to choose for them.

Intrigued by the stark contrast of outlook and expectation between these two patients, I excuse myself from the clinic to find them in another room awaiting further tests. After some general discussion about their respective health issues, I ask them individually what the doctor's position should be in their decision-making. They unanimously maintain that one of the doctor's main roles is to fully inform them. However, the first patient feels that a doctor's experience warrants choosing for her, while the second patient comments that he is tired of having to do what doctors tell him to do. It became clear to me that there is no singular view of what autonomy means. One interpretation, as expressed by the second patient and championed by the philosopher Isaiah Berlin, incorporates a complete freedom from external constraint; an utter self-sufficiency and responsibility for all aspects of life. Notably, in upholding such a stance, autonomy simultaneously becomes a duty of sorts; a patient who chooses that they trust their doctor enough to make or strongly influence their decision for them cannot be viewed as autonomous[4]. Alternatively, a view of autonomy centred on freedom of choice rather than on complete independence allows a patient like the former to maintain her autonomy while passing some responsibility of the decision-making process to the doctor. The American philosopher Gerald Dworkin argues that autonomy does not require independence for its own sake: provided the patient is fully informed and engages actively in the decision-making process they may still be autonomous while being receptive to, or even reliant on, the opinion of the doctor[4,7].

Therefore, for the doctor to respect this patient's autonomy, perhaps he/she has to accept that this patient does not wish to make the decision solely by themselves (provided they are fully informed and their judgement is a rational one), i.e. an autonomous delegation of choice. Furthermore, accepting this is also crucial for the doctor to act beneficently in this situation. To quote Dworkin: "autonomy is important, but so is the capacity for sympathetic identification with others. … [A]lthough it is important to respect the autonomy of others, it is also important to respect their welfare, or their liberty, or their rationality"[8].

In conclusion, I believe it is time to end the frustrating strife to abolish all the dwindling traces of paternalism from modern medicine at the expense of our patients' welfare. It is time to stop sacrificing beneficence for the respect of a universal notion of what patient autonomy should be and instead to carefully determine each patient's view of what their autonomy entails, fully inform them so they are in a position to exercise this autonomy, and mould our relationship with them around this deepened understanding. If we can achieve this – a respect for autonomy tailored to each patient who walks through the clinic door, be it either of the two patients on that November morning or anyone in between – an "overemphasis" of autonomy can do nothing but good.

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The Pavlik Harness as Treatment for Developmental Dysplasia of the Hip

Winning Article, TSMJ Paediatrics Essay Prize, 2013

Catherine Quinn, 4th Year Physiotherapy, TCD

> **"The Pavlik harness has been shown to be extremely successful in the treatment of developmental dysplasia of the hip (DDH). Risk factors for failure and complications have been identified and can therefore be minimized."**

Developmental dysplasia/dislocation of the hip (DDH) is a congenital condition that represents a number of anatomical abnormalities where the hip joint is not fully formed at birth. The femoral head is not held firmly in the acetabulum. It can be caused by the acetabulum being too shallow and/or the hip ligaments being lax. This means that it can range from slight ligament laxity to complete dislocation. Newborns are screened for DDH, as left untreated it can lead to abnormal gait, pain and osteoarthritis in early adulthood[1]. It is more common in girls, especially first-borns, and tends to run in families[2]. There is no definite cause; it may be a reaction to the mother's hormones during pregnancy, due to a tight uterus that does not allow foetal movement, or being born breech. It is often graded by Graf classification, ranging from I to IV, with IV being the most severe form of dislocation. The Pavlik harness is the most common harness used to treat DDH[3].

Ultrasound is reported as the most sensitive method for detecting hip abnormalities in infants[4], followed by clinical examination using the Barlow and Ortolani manoeuvres. Both tests are performed with the hip and knee bent to 90 degrees. Barlow's test is performed by adducting the hip and applying a downward pressure. The feeling of the femoral head slipping out of the socket posterolaterally indicates a positive result. The Ortolani manoeuvre involves gently abducting the hip and pulling the femoral head anteriorly. An audible and palpable clunk demonstrates a positive result. However, a study by O'Grady et al.[5] found that 84% of the units they included in their study (n=19) in the Republic of Ireland relied on radiographs for diagnosis. Only 37% had access to hip ultrasounds and only 42% of the units included in their study had formal DDH screening. This study highlights the fact that the two most effective methods of diagnosis are not routinely being used in Ireland today. Another study states that almost all authors agree that Ortolani-

positive hips should be treated and all Barlow-positive hips should be treated[3]. It can be difficult to decide whether or not slightly unstable hips should be treated with the Pavlik harness, as it puts healthy or almost healthy hips at a higher risk of complications, with avascular necrosis (AVN) being reported in healthy hips.

The Pavlik harness is the preferred treatment for DDH. First presented by Arnold Pavlik in Prague in 1946[6], the harness prevents extension and adduction but allows for safe movement. The weight of the limb generates a force that repositions the femoral head. Sleep appears to be an essential factor in repositioning, as the reduced muscle tone allows the bone to move[7]. The abduction caused by the harness stretches the adductor muscles, and the hip flexion caused by the harness helps to shift the head of the femur posteriorly into the acetabulum from its dislocated anterior position. In a newborn with an Ortolani-positive or Barlow-positive hip, stability is usually seen on clinical examination after two to three weeks. It is often recommended to continue wearing the harness for six to eight weeks to maintain the integrity of the joint[3]. Van der Sluijs showed that the Pavlik harness can be worn for up to 32 weeks before stability is reached, without resulting in AVN[8]. In an infant over three or four months, the hip must be monitored clinically and ultrasonographically, with at least three weeks of continuation of treatment recommended when stability is reached. The child is then gradually weaned off the harness for a few hours each day. If no reduction is seen after three weeks in the Pavlik harness, an attempt may be made to carry out a closed reduction after a period of skin traction. If a closed reduction proves unsuccessful, an open reduction must be performed. After a reduction has been obtained, the child should be placed in a spica cast in 95 degrees of hip flexion and 55 to 65 degrees of abduction.

The Pavlik harness has been reported with various success rates. Walton et al. report a 100% success rate of the Pavlik harness in treatment of subluxatable hips, although they report a 100% failure rate of hips that were irreducible on ultrasound scan on initial assessment[9]. Peled et al. report a 95.8% success rate for the Pavlik harness in treating Graf Type III hips and 61.5% in Graf Type IV hips. Similarly, Mostert et al. report relocating 97% Graf Type III hips and 50% of Type IV10, and Malkawi recorded a 100% success rate in 699 hips[11]. However Atalar et al. report only 58% successful reduction in hips graded Graf IIc or more severe with the Pavlik harness[12], and Wilkinson et al. report that 30% of their 43 hips graded Graf III or IV required further treatment[13]. However, it should be noted that the studies with high success rates report early diagnosis and intervention, and the studies with lower success rates commenced treatment later. Therefore, the Pavlik harness has been accepted as an effective treatment for Graf III and some Graf IV hips, with early intervention increasing the effectiveness of treatment. According to Vitale and Skaggs, patients under six months of age have a better outcome[14].

Therefore, if early diagnosis and treatment is essential to outcome of treatment, it is imperative that the clinician is aware of predisposing factors that lessen the effect of the Pavlik harness or are predictive of failure. A study found that there was no association between success and gender (although this study of 23 patients included only two boys), side of pathology or findings on clinical examination[12]. However it did support previous findings, stating that hips in which treatment commenced before the patient was seven weeks of age had a higher success rate than patients of eight weeks or over. They also found that the less severe the dislocation, the better the results, with hips graded IIc, IId and III resolving better than hips graded IV. Patients presenting with these factors should be monitored extremely closely to identify failure of treatment so that another appropriate treatment can be started as soon as possible.

As with many treatment techniques, the use of the Pavlik harness has complications and risks. The most serious risk is AVN. AVN is caused by forced attempts at reduction, especially in

abduction, which can damage the blood supply to the growing head of femur and therefore allow the bone tissue to die. Pap et al. reflected on patients treated between 1974 and 1982, a period in which ultrasonography was not routinely used to screen DDH and therefore many patients were unnecessarily treated[15]. The overall occurrence of AVN in this study was 11.7%. They found that there was a strong relationship between age at the start of treatment and AVN forming on the dysplastic side, the younger hips being less likely to develop necrosis. This study also showed that AVN can form in the healthy contralateral hip. There was no relationship between age at the start of treatment and AVN in the healthy hip, but it was linked to a prolonged length of treatment. However it should be noted that no AVN severe enough to cause substantial damage (Tönnis grade III) was found on the healthy side. Suzuki et al. reported an 8% incidence of AVN in 19967. They found no connection between age and occurrence of AVN; however, a close relationship was seen between the type of dislocation and the presence of AVN, the more severely dislocated hips being more likely to develop AVN. Reduction usually occurs during sleep due to the weight of the limbs and decreased tone, but as extreme abduction can decrease blood supply, the author recommended placing pillows under the legs while sleeping to prevent this.

Another possible complication is the development of femoral nerve palsy. This is thought to be caused by high degrees of hip flexion compressing the femoral nerve under the inguinal ligament. A study by Murnaghan et al. reports a 2.5% prevalence of femoral nerve palsy in their study of their 1218 patients[16]. 87% of the cases were diagnosed in the first week of treatment. All babies had complete return of femoral nerve function, with no reports of permanent damage. It appears that larger or heavier babies are at an increased risk of femoral nerve palsy, as are older babies, but no relationship was found between sex, bilaterality, ethnicity or birth weight. Interestingly, the success rate of the Pavlik harness in the palsy group was 47% compared to 94% in the control group, with the impact of femoral nerve palsy being greater on hips that were Ortolani-positive or hips with a fixed dislocation. If the palsy lasted more than three days there was a 30% success rate for the Pavlik harness, which shows the importance of checking the quadriceps at follow-up visits. The author advised temporarily removing the harness until the palsy resolves, but not completely abandoning it.

As mentioned previously, it is essential that healthcare professionals monitor the hips for development and complications. They must endeavour to do all that they can to ensure the harness is used correctly. However, realistically, there is only so much that can be done at appointments, so the responsibility lies with the parents. Hassan et al. studied the compliance of parents with the Pavlik harness[17]. The study included a wide variety of mothers, with ages ranging from 20 to 42 years old, some with a university level of education (being the majority group at 38.12%), some with college level or high school education and the minority having below high school level (9.51%). Upon first seeing the harness, without seeing its application, 33.8% reported it would be easy to use, 45% said difficult and 21.3% said complex. However, after watching it being applied and trying it for themselves this changed to 96.25% reporting it as easy and 3.75% calling it difficult. No relationship was noted between the mothers' level of education and their emotional reaction to the harness. 94.37% reported following the physicians advice strictly as they believed their child had a serious condition, resulting in an average duration of 6 to 16 weeks of treatment. The 3.12% who reported removing the harness for portions of the day (as they did not view the condition as serious) had an average treatment duration of 12 to 18 weeks. Again there was no relationship between the parents' educational level and compliance. 96.25% reported receiving adequate information about application and use of the harness, although 48.1% would have preferred to receive a leaflet with written instructions, again with no link to education. 22.66% reported difficulty reapplying the harness after bathing in

the first week. 61.87% were concerned to leave the child a week without bathing, and 88.8% had significant emotional difficulties with the child being uncomfortable. This study shows that most parents are compliant with the treatment, but highlights factors that influence compliance. The healthcare provider must emphasise the importance of using the harness exactly as prescribed to ensure the best outcome for the child. They must ensure that the parents take the condition seriously as they govern the treatment. This study also reminds the reader that use of the Pavlik harness can be emotional and distressing for parents who are anxious about their new baby. They must be supported, encouraged and educated about the treatment, and provided with as much information as possible.

The Pavlik harness has been shown to be extremely successful in the treatment of DDH. Risk factors for failure and complications have been identified and can therefore be minimized. From the evidence given above, it seems that early diagnosis and intervention, along with regular check-ups, are crucial to both treatment effectiveness and avoidance of AVN. It must be remembered that a diagnosis of DDH may be traumatic for parents, who must be educated on the successfulness of the harness and certain complications for which to look out. With co-operation between the parents and the medical team, the Pavlik harness can avoid a lifetime of hip problems and give a child a normal life.

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Neurosurgical Treatment of Moyamoya Disease: Bypass Surgery for the Brain

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> "Currently, no medical treatment exists to prevent the progression of moyamoya disease, and neurosurgical treatment is the accepted means of primary treatment."

Abstract

Moyamoya disease leads to disruption of the blood supply to the brain and affects populations around the world; the highest prevalence is reported in the Japanese population. Symptoms of this disease may be due to ischaemia, including transient ischaemic attacks and stroke, or due to haemorrhage from abnormal and weak collateral vessels, leading to loss of consciousness or acute-onset headache. This disease is progressive and cannot be adequately treated medically, requiring neurosurgical intervention for effective treatment. Surgical treatment seeks to increase blood flow to the brain and may be done either by the direct anastomosis of the superficial temporal artery and middle cerebral artery or by a variety of indirect approaches that act to increase collateral blood flow to the brain. Although no standard treatment exists for this disease, many surgical interventions and combinations of interventions have been used with successful outcomes.

Introduction

Moyamoya disease (MMD) is a condition affecting the vasculature supplying the brain, predisposing individuals affected by this disease to stroke. This disease leads to stenosis of the internal carotid arteries (ICA) within the skull, which normally act to supply the brain with the rich blood supply it needs to function[1,2]. The narrowing of the carotid vessels seen in MMD is unlike that seen with atherosclerosis, a more common cause of stroke, and perhaps the most striking difference between these two causes of stroke is the age groups which are affected: while the majority of strokes affect individuals greater than 65 years of age, MMD may lead to stroke in children younger than 3 years old[1,3]. This article will look at how MMD affects both children and adults, how the presentation of this disease differs significantly between children and adults and, lastly, what treatments are available for this disease.

What Causes Moyamoya and Who Does it Affect?

もやもや

Moyamoya is a Japanese word, written above in hiragana, and means puff of smoke. The name originates from the characteristic hazy appearance of angiograms seen in patients with this disease, a result of the growth of collateral vessels that seek to compensate for the limited blood supply provided by the carotid arteries (Figure 1)[1,4,5].

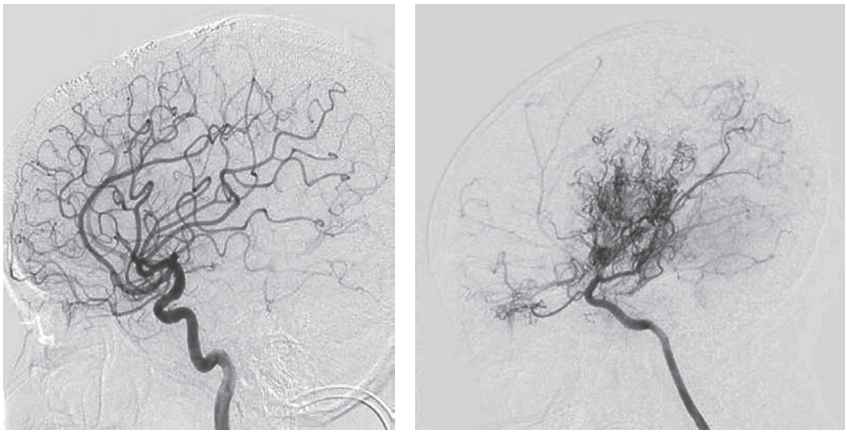

Figure 1. *(Left) Lateral view angiogram of the internal carotid artery (ICA) showing normal perfusion of the brain. (Right) Angiogram of a patient with moyamoya disease showing the "puff-of-smoke" appearance which can be seen in patients with this disease[1].*

First described in Japan in 1957, the cause of MMD remains unclear but it is believed that environmental and genetic factors play a role in its development. Familial cases of MMD have been reported in Asian populations and risk increases to 30 to 40 times that of sporadic cases in first- and second-degree family members of these patients. There also appears to be an association between MMD and genetic disorders such as Down syndrome and neurofibromatosis. However, the majority of presentations of MMD occur sporadically[6-8]. Pathological changes underlying the stenosis of the major vessels supplying the brain do not involve an inflammatory reaction but instead are thought to centre around smooth muscle cell proliferation[9,10]. The brain tissue, in response to the decline in blood supply, stimulates angiogenic cascades, leading to the characteristic collateral vessel growth seen in MMD[2]. Increased blood flow across these weaker collateral vessels leads to high haemodynamic stress, which in turn can cause microaneurysm formation and vessel wall necrosis, changes which are thought to predispose to the haemorrhage seen in this disease[10,11].

Prognosis

MMD is known to be progressive, with rates of morbidity in untreated individuals as high as 65%-70%[2,12,13]. Ischaemia in MMD may result in the loss of the ability to speak, blindness due to cerebral damage and, in long-standing cases, a persistent vegetative state[5]. In cases presenting with haemorrhage, the ability to recover successfully is drastically reduced with each subsequent haemorrhage. After the first episode of haemorrhage, successful recovery is reported at around 45%; after a second bleed, this falls to about 21%. Mortality after the first bleed has been reported at 7%; after a second haemorrhage, this mortality rose to a startling 29%[14]. The high morbidity and mortality associated with both ischaemic and haemorrhagic presentations of MMD highlight the need for effective control of this disease.

Incidence

MMD was originally thought to be a disease solely affecting Japanese and Korean populations; however, the disease has now been described in many other countries, albeit at a much lower incidence[4,10,15]. The prevalence of MMD in Japan is around 3 per 100,000 people, whereas in Europe and the United States, a lower prevalence is observed at about 0.3 and 0.09 per 100,000, respectively[10,16]. Females are more frequently affected by this disease than males, with reported ratios ranging from 1.5 to 2 females for every male affected[10,16]. The disease sees two peaks in incidence, one in children at 5 years of age and again in adults in their mid-40s[1,16]. It is currently the most common cerebrovascular disease affecting the paediatric population in Japan and accounts for 10% to 20% of arterial infarction in children globally[9,16].

Recognising Moyamoya

Presentation

The presentation of moyamoya corresponds with the pathological changes observed. Stenosis causes ischaemia which leads to transient ischaemic attack (TIA), stroke and seizures[1]. In children, episodes of crying and hyperventilation can lead to decreased partial pressure of CO_2 in blood and reflex physiological cerebral vasoconstriction, precipitating symptoms of stenosis. This, in addition to the already diminished blood flow in MMD, can lead to ischaemic events[17]. Patients with MMD may experience persistent headaches, thought to arise from irritation caused by the dilation of collateral vessels within the meninges[1,5]. The cerebral haemorrhage observed in MMD may present with an acute-onset headache but may also lead to loss of consciousness or motor control disturbance[5]. Haemorrhage occurs about 7 times more frequently in adults than in children and presentations due to haemorrhage make up about 66% of MMD cases in adults. Within adults, haemorrhage occurs more commonly in Asian populations than American ones[1,5]. Ischaemic presentations make up about 69% of those seen in children less than 10 years of age[5]. Unfortunately, completed stroke occurs more often in children and is thought to be due to difficulty in reporting symptoms because of limited verbal skills in younger age groups[1]. Children affected by this disease may also experience intellectual disability and reduced IQ[5]. Occasionally, MMD may be identified incidentally in asymptomatic individuals; in such patients, careful monitoring should be undertaken to identify progression of the disease and treatment should be considered[5,18].

Diagnosis

The criteria for diagnosing MMD are defined by the Research Committee on Moyamoya Disease of the Ministry of Health and Welfare in Japan. Based on these guidelines, MMD involves bilateral occlusive change in the terminal ICA and/or proximal portions of the anterior or middle cerebral arteries (MCA), abnormal vascular networks in the region of the occluded vessels and the absence of systemic disorders which might account for these changes[2,10]. The diagnosis of MMD can be carried out using a variety of imaging techniques. Computed tomography (CT) may be used to show haemorrhage in MMD, while magnetic resonance imaging (MRI) may be useful in identifying areas of ischaemia. Traditional angiography and the less invasive magnetic resonance angiography are useful in identifying stenosis across the major vessels supplying the brain[1,19,20]. Cerebral blood flow may be analysed using single-photon emission computed tomography (SPECT) or xenon-enhanced CT scanning, the latter of which could have the potential to be used as a tool for determining stroke risk in patients with MMD as well as the likelihood of ischaemia following revascularization[19,21-23].

Clinical Points

1. Moyamoya disease is a progressive disease and an important cause of stroke in children and young adults.
2. Moyamoya disease most often occurs sporadically.
3. Children with moyamoya disease often present with signs of ischaemia while adults present more commonly with signs of haemorrhage.
4. Neurosurgical intervention by either direct or indirect revascularization techniques is accepted as the primary means of treatment for this disease.
5. Both direct and indirect revascularization techniques halt progression of the disease and effectively reduce morbidity and mortality associated with this disease.
6. No randomised control trials have yet been carried out to determine a standard treatment course.

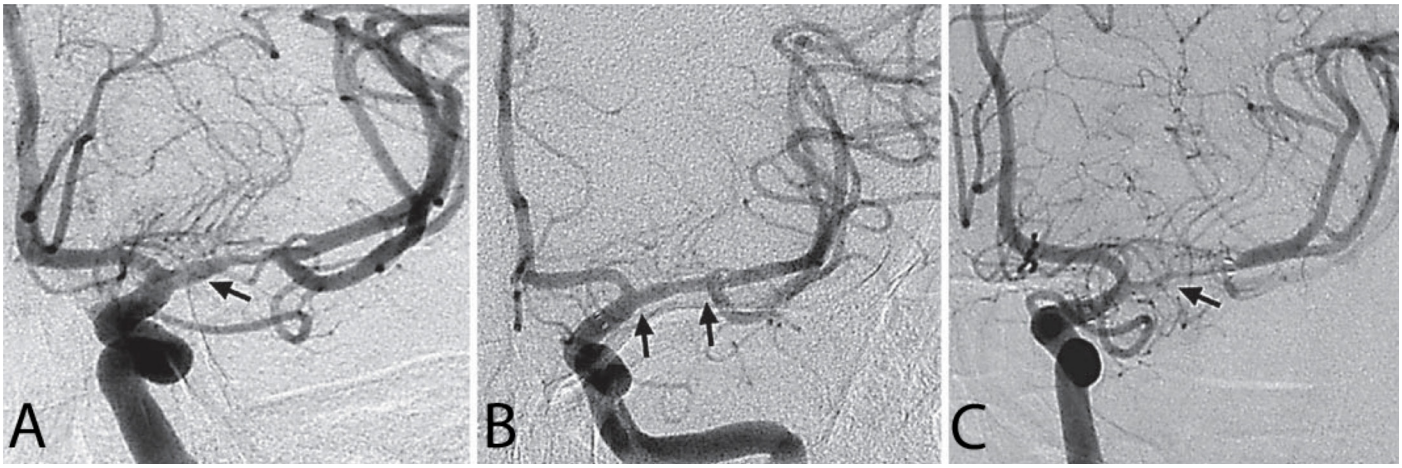

*Figure 2. **(A)** Anteroposterior angiogram of the left ICA showing stenosis of the MCA (arrow). **(B)** Immediately after angioplasty and stenting of the patient's MCA, we see an increased patency of the MCA (arrows). **(C)** 6 months after stenting, re-stenosis of the MCA is noted (arrow)[26]. Reproduced with permission from S. Karger AG Publishers, Basel (Switzerland).*

Treating Moyamoya

Currently, no medical treatment exists to prevent the progression of MMD, and neurosurgical treatment is the accepted means of primary treatment[1,17]. The neurosurgical approach to treatment of this disease aims to increase blood flow in areas of the brain where flow has been compromised, in order to avoid the ischaemia occurring due to vessel stenosis, and also theoretically reduce the strain on collateral vessels in order to reduce haemorrhage and headache. However, whether or not haemorrhagic disease fully responds to revascularization is still up for debate[24,25]. It is interesting to note that accepted treatments for stenosis in atherosclerotic disease, such as balloon angioplasty and stent placement, fail to effectively treat the hypoperfusion seen in MMD (Figure 2)[26]. Most currently accepted surgical interventions for MMD take advantage of the fact that the external branch of the carotid artery is not affected by the disease[1].

There are two general approaches to surgical treatment: direct revascularization and indirect revascularization.

Direct Revascularization

Direct neurosurgical intervention involves joining a branch of the superficial temporal artery (STA), a branch of the unaffected external carotid artery, to the MCA, a major cerebral blood vessel which arises from the now partially occluded ICA[1,2,27]. This procedure was first performed in the 1970s and is similar to a coronary artery bypass graft (CABG) operation where a patent vessel is used to bypass occluded coronary vessels to restore adequate blood flow to the myocardium.

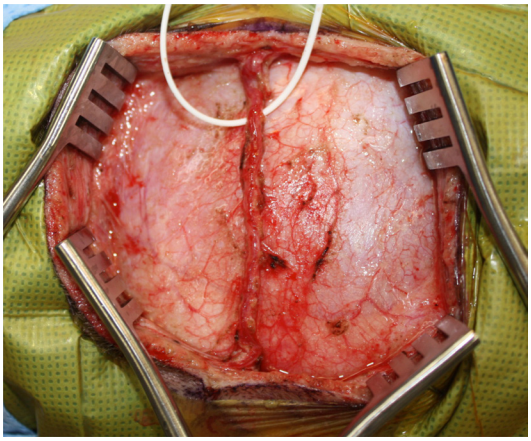

Figure 3. Intraoperative photo showing retraction of the scalp and isolation of the superficial temporal artery (STA). This initial step is similar for STA-MCA anastomosis, encephaloduroarteriosynangiosis and pial synangiosis. Photo provided by Dr Jodi Smith, Associate Professor of Neurological Surgery, Indiana University School of Medicine.

During the neurosurgical approach, the first step involves locating a branch of the STA using Doppler ultrasound followed by careful dissection to expose this vessel (Figure 3). The STA is then moved out of the operative field and a craniotomy delicately performed to avoid damaging the middle meningeal artery: a vessel within the skull that tends to supply important collateral vessels to the brain of patients with MMD. After the craniotomy, the dura (the thick outermost component of the meninges) is opened,

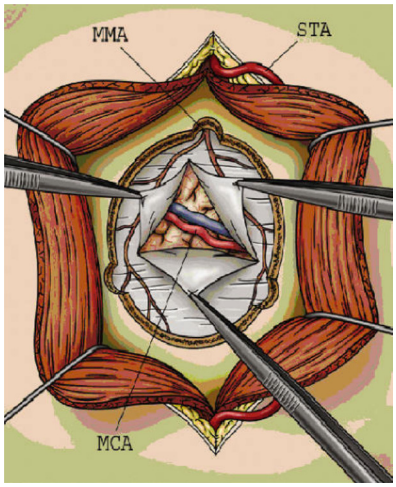

Figure 4. Figure showing the superficial temporal artery (STA) out of the operative field, the preserved middle meningeal artery (MMA) as it courses below the skull, and the middle cerebral artery (MCA) where the dura, pictured here held by forceps, has been removed[10].

exposing the brain's surface and vasculature, now visible through the thinner arachnoid membrane of the meninges. At this point a large-diameter artery is selected for the anastomosis (Figure 4). After identification of an appropriate artery, the arachnoid is opened around the vessel and a segment of the artery is prepared for the anastomosis. Clips are used to prevent bleeding during anastomosis and the previously dissected STA is incised from its distal end and adjoined to the MCA via a longitudinal incision made in the side of this artery (Figure 5)[1,4,10].

Problems arise with this approach in children as their superficial temporal arteries are very small, making the procedure technically difficult to perform[1,10]. Direct techniques are also made more difficult in children due to a higher risk of thrombosis in these smaller vessels. In general, bypass also carries a higher risk of detrimental post-operative haemorrhage[4]. Another complication associated with STA-MCA anastomosis is cerebral hyperperfusion resulting from the sudden increase in blood flow across the MCA. This leads to symptoms such as severe headache or focal neurologic deficits. Fortunately, however, though these patients may appear to show signs of ischaemia, they often make a full recovery from this complication, and it may be avoided altogether with careful management perioperatively[28,29]. Direct STA-MCA anastomosis currently remains a valuable means of treatment for MMD1.

Indirect Revascularization

Indirect treatment options also act to bypass the occluded internal carotid artery supply via the external carotid blood supply, but these approaches do not directly join vessels. Instead, indirect approaches depend on new vessel growth by either tissue or vasculature supplied by the external carotid artery to areas of ischaemia within the brain. These approaches include encephalomyosynangiosis (EMS), encephaloduroarteriosynangiosis (EDAS), pial synangiosis and multiple cranial burr holes, to name a few[1,4,10].

Encephalomyosynangiosis, like direct STA-MCA anastomosis, was first used in the 1970s for the treatment of MMD. This procedure involves suturing the well-vascularised temporalis muscle to the surface of the dura. This may lead to collateral vessel formation should angiogenesis be successful. Disadvantages of this procedure include pressure on the brain because of the additional tissue within the skull, as well as an increased incidence of seizures[10].

During encephaloduroarteriosynangiosis, instead of transecting the STA as in the STA-MCA

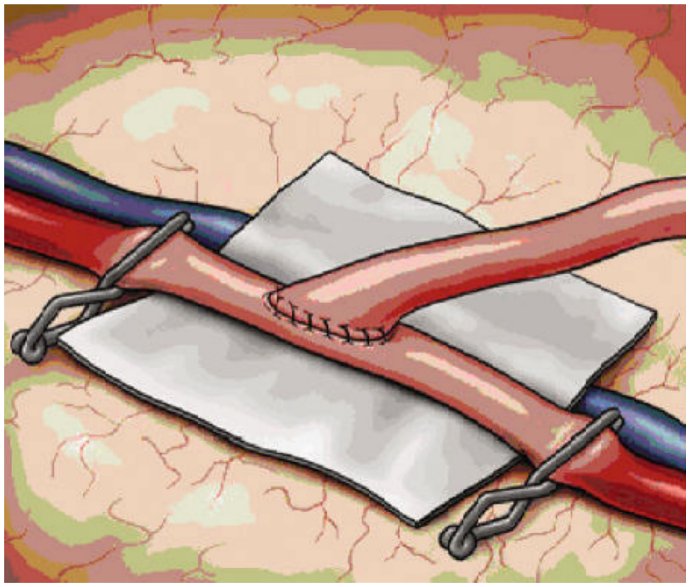

Figure 5. The MCA is represented here in this diagram in red with clips attached, coursing parallel to an adjacent vein in blue. In the centre of the diagram we see the STA anastomosed to the MCA[10].

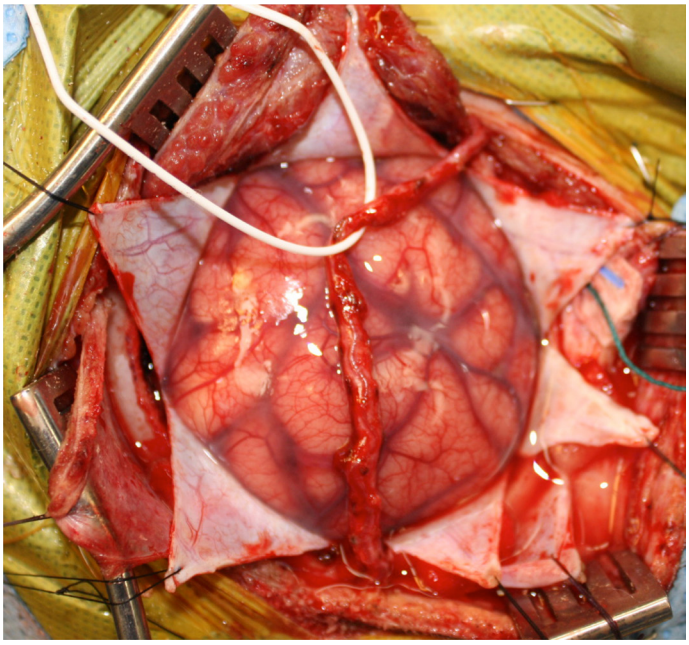

Figure 6. *Intraoperative photo demonstrating a pial synangiosis being performed. The scalp is retracted, bone flap removed and dura opened in a stellate fashion exposing the brain. The branch of the STA to be sewn to the pia can be seen running across the operative field. Photo provided by Dr Jodi Smith, Associate Professor of Neurological Surgery, Indiana University School of Medicine.*

anastomosis, both its attachments are preserved. A craniotomy is performed and the identified vessel is sewn to the dura, then the bone flap is carefully replaced so as to allow patency of the vessel as it passes from scalp to below the skull[4,10]. A pial synangiosis takes this one step further and instead of attaching the artery to the dura, the dura and arachnoid are opened and the vessel sutured directly to the brain surface (Figure 6) before the bone flap is replaced (Figure 7)[10,30]. A follow-up study of 143 patients receiving pial synangiosis for the treatment of MMD found this procedure to be effective in reducing stroke and transient ischaemic attacks and effectively halting the normal course of clinical deterioration[30].

The placement of multiple burr holes is the least technical of these procedures but can provide effective blood vessel growth into targeted localised areas by creating a conduit along which collateral vessel growth may occur[4]. Additional procedures are available for indirect revascularization involving various modifications to these techniques and combinations chosen to target an individual's specific needs[4,10].

Because connections between vessels are not directly made during indirect techniques, increases in blood flow are not seen immediately after surgery. Long-term, however, there appears to be no advantage in using direct techniques over indirect techniques[1,2]. There is conflicting evidence regarding the efficacy of indirect techniques in older patients, with some reporting slightly less benefit from indirect techniques in these patients as compared to paediatric populations[2,10,31,32]. It is proposed that this may be a function of a decline in angiogenic potential with age[10]. Direct arterial bypass to regions other than those supplied by the MCA is often not technically feasible and in these situations, indirect procedures are often favored. Indirect techniques can also be performed faster, reducing complications associated with general anaesthetic[10].

Retrospective analysis of 410 paediatric patients treated for MMD at a single institution supported the early identification and neurosurgical treatment of MMD in order to achieve favourable outcomes[33]. However, there is no standardised approach to MMD

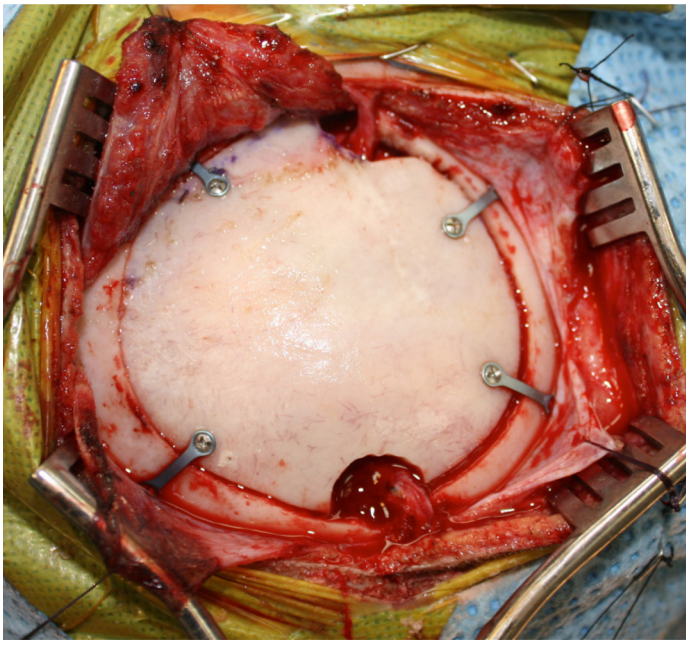

Figure 7. *Intraoperative photo demonstrating the replacement of the bone flap after a pial synangiosis has been carried out. Notice the bone flap has been altered to allow the STA to flow freely beneath the skull and out again. Photo provided by Dr Jodi Smith, Associate Professor of Neurological Surgery, Indiana University School of Medicine.*

and randomised control studies comparing the various surgical techniques do not yet exist[17].

Conclusion

Moyamoya disease is an important cause of stroke in children and young adults and should be suspected in any individual presenting with symptoms of stroke or TIA at a young age. A standardised approach to the treatment of MMD does not exist but direct and indirect revascularization techniques appear to have comparable efficacy in treating this disease. Research is still needed to address whether the haemorrhage seen in MMD can be effectively treated with current revascularization techniques and whether indirect revascularization is as valuable as direct revascularization techniques in adult populations.

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To Never Know Heartbreak – Fetal Cardiac Intervention to Treat Hypoplastic Left Heart Syndrome

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> "Fetal cardiac intervention modifies the course of cardiac growth, function and/or development in utero sufficiently to alter the postnatal outcome."

Abstract

Within the last three decades, no congenital heart defect has undergone a more dramatic change in management and outcome than hypoplastic left heart syndrome (HLHS), which is an invariably lethal heart defect without treatment. During this time, we have seen a change from a fatal diagnosis to a successful treatment where 70% of those diagnosed with HLHS will reach adulthood, albeit with a single ventricle system or "Fontan circulation". As the pathogenesis of HLHS is being elucidated, fetal cardiac intervention (FCI) is becoming a real possibility. It has been hypothesised that FCI could potentially halt the development of HLHS and consequently create a biventricular system. Two predominant research groups have shown promising results and have achieved biventricular circulation as a postnatal outcome in 30–67% of neonates. It is believed that with further research, improved instrumentation and more advanced imaging, we will see significant progress not only in treatment of HLHS but of congenital cardiac defects overall.

Introduction

Congenital heart disease is the most common inborn defect, occurring in 19/1000 live births[1]. Over 20 years ago, the idea of fetal cardiac intervention (FCI) was put forth to treat congenital heart defects (CHD). The idea arose due to the observations that some forms of cardiac malformations progressed in severity as the pregnancy progressed[2]. Fetal cardiac intervention modifies the course of cardiac growth, function and/or development *in utero* sufficiently to alter the postnatal outcome[3]. There is also evidence that prenatal intervention may allow the fetus to recover in the supportive environment found *in utero* that encourages enhanced wound healing and myocyte proliferation[4,5]. Fetal cardiac intervention is most effective in cases where intervention may alter the evolution of the condition or if the fetus is at risk of

death. It is therefore indicated if the severity of postnatal disease may be substantially reduced[3] (Table 1 includes examples of such conditions in which FCI is feasible).

Of the CHDs, none has been researched more for FCI than hypoplastic left heart syndrome (HLHS). This syndrome is a lethal congenital heart defect that is associated with obstruction to left ventricular outflow[6], leaving the left heart complex underdeveloped[7]. The degree of hypoplasia is proportional to the severity of obstruction, except in circumstances where there is an alternative route of blood flow such as with a ventricular septal defect. If the hypoplasia is severe, the left ventricle (LV) cannot support systemic circulation[6], which can lead to development of right heart compression and severe hydrops *in utero*[8] or demise soon after birth.

Fetal echocardiography has allowed for prenatal diagnosis and assessment of HLHS, demonstrating that LV hypoplasia severity evolves throughout gestation. This implies that normal cardiac morphogenesis requires blood-flow-directed remodelling in addition to intrinsic patterning. Therefore, abnormal blood-flow streaming may lead to HLHS[6].

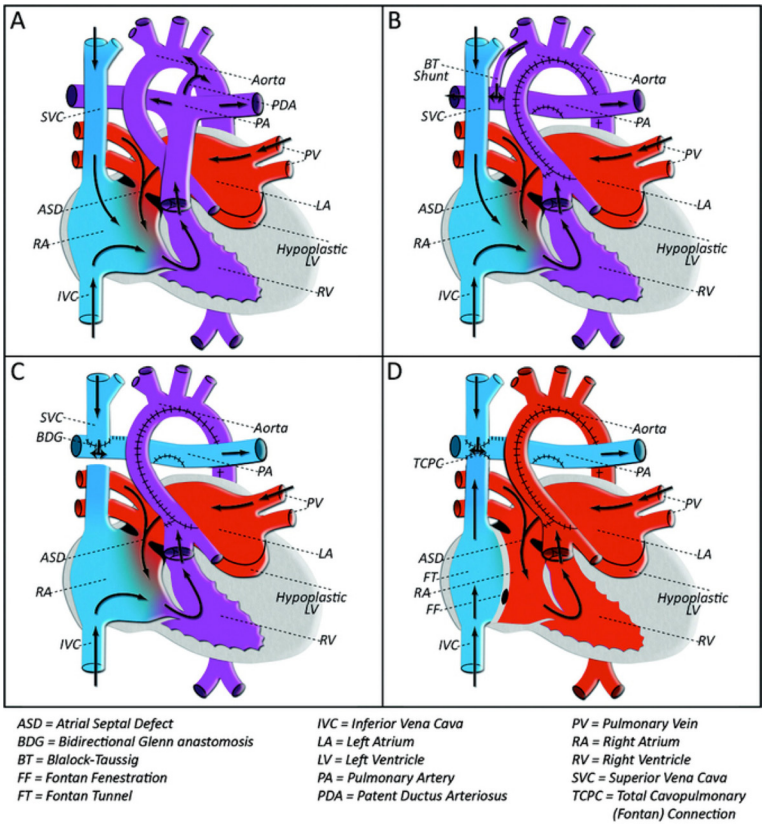

Figure 1. *3-stage palliative repair of a neonate diagnosed with HLHS. Heart with HLHS (A), Norwood Procedure (B),Glenn Operation (C), Fontan Procedure (D)[26].*

Current Treatment for HLHS

Newborns with HLHS (Figure 1A) may be asymptomatic, but as soon as the ductus arteriosus closes they become severely ill and, without treatment, almost invariably die[7]. The current treatment of HLHS that allows a 70% survival rate into adulthood includes a three-stage palliation procedure beginning with the Norwood procedure shortly after birth[9]. This procedure converts the right ventricle into the main systemic ventricle while

Table 1. Congenital Cardiovascular Anomalies Potentially Amenable to FCI

Condition	Actual or Possible FCI
Fetal tachycardia with hydrops	Maternal antiarrhythmic pharmacology
Structural anomalies causing hydrops	Maternal digoxin
Congenital heart block	Pacemaker
Severe Ebstein malformation	Tricuspid valve repair
Severe congenital MR or AS	Balloon or surgical valvuloplasty
HLHS with intact atrial septum	Creation of atrial septal defect
Evolving hypoplastic right heart	Pulmonary valve perforation and dilation
Premature closure of ductus arteriosus	Ductal stenting
Absent pulmonary valve syndrome	Pulmonary arterioplasty

McElhinney et al.[3]

Clinical Points

1. Hypoplastic left heart syndrome (HLHS) is a lethal congenital heart abnormality that usually occurs due to left ventricular outflow obstruction.

2. In the past 30 years, the only treatment option for HLHS has been a sequence of complex open-heart operations that lead to a univentricular "Fontan circulation".

3. Potential long-term outcomes of Fontan circulation include increases in the development of arrhythmias and coagulopathies and poor exercise tolerance' leading to heart failure and neurological disability.

4. Fetal cardiac intervention is theorised to perhaps prevent the development of HLHS by making the stenotic aortic valve patent and allowing fetal cardiac flow dynamics to run their natural course.

5. Postnatal outcomes of FCI and aortic valvuloplasty that resulted in biventricular circulation for HLHS vary between 30 and 67%.

6. Advancement in instrumentation and imaging will lead to improved outcomes of FCI and a potential to prevent HLHS before it manifests.

the aorta and pulmonary trunk are joined together[10]. The pulmonary arteries are cut away from the pulmonary trunk and a Blalock Taussig (BT) shunt is created to allow blood flow from the innominate artery to the pulmonary arteries (Figure 1B)[11].

Six months after the Norwood procedure, the bi-directional Glenn operation[10] (Figure 1C) occurs. The superior vena cava (SVC) is connected to the pulmonary artery and the BT shunt is disconnected. This will send blood directly to the lungs without its having to pass through the univentricle.

Lastly, the Fontan procedure occurs between 18-36 months and connects the inferior vena cava (IVC) to the pulmonary artery. Consequently, all the systemic venous blood will flow directly into the lungs[10,12], become oxygenated, then enter the univentricle to be distributed to the systemic blood supply (Figure 1D).

This procedure has demonstrated success, but numerous complications can arise. Following the Norwood procedure, supra-ventricular tachycardia occurs in about 15% of cases, infection affects about 10% and bleeding and coagulopathy are almost universal. Lastly, seizures occur in 17-22% of patients and are associated with developmental delay[9].

Long-term complications can occur as well. As the original Fontan operation was performed in 1971, the first cohorts of adult patients are now being looked at for evidence of long-term complications. Arrhythmias, thrombosis, protein-losing enteropathy, and increasing exercise intolerance leading to heart failure after 15-30 years are a few examples[12]. Once this ventricle fails, transplant is the only option[2]. Van den Bosch et al. studied a cohort of patients who underwent the Fontan procedure with a mean follow up period of 15 years. They demonstrated that 28% had died at a mean age of 10 years old while 58% of patients underwent a reoperation to revise the Fontan connection. Supraventricular tachycardia was observed with an increased incidence of arrhythmias in 56% of patients, and finally, 25% experienced thromboembolic events that resulted in fatalities of three patients[12]. It should be noted that this three-stage palliation procedure is life-saving for many neonates who would otherwise succumb to their condition. These results reflect a procedure that took place about 30 years ago[6]. Since then, it can be postulated that experience and new research have improved the technicalities of the procedure and that future cohorts may show improved long-term outcomes. However, it can also be inferred that there is room for an improved technique in treating HLHS and perhaps an intrauterine approach (FCI) will prove to be just that.

Fetal Cardiac Intervention

Despite improved surgical outcomes achieved with the three-stage palliation procedure, the question

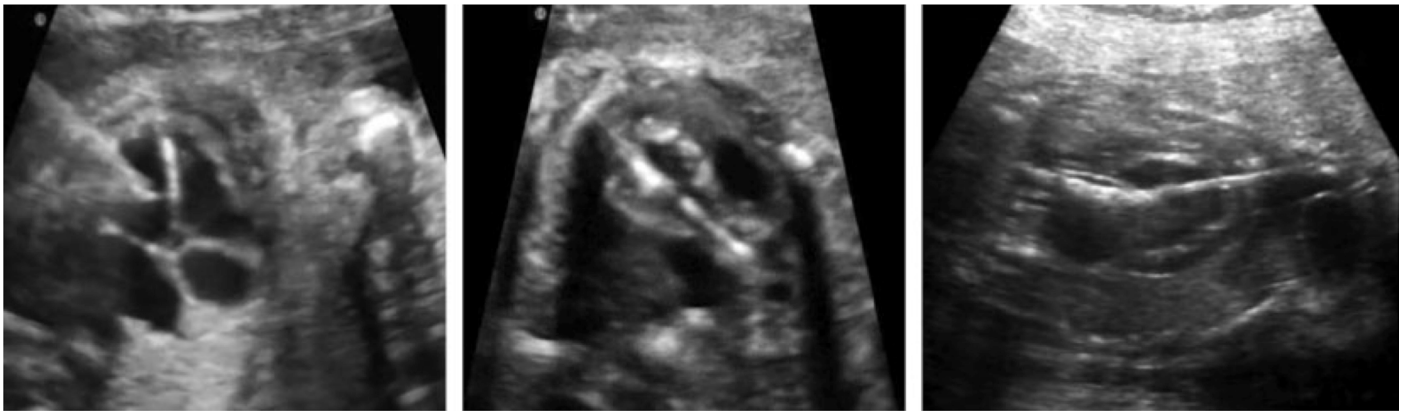

Figure 2. *Ultrasound images in the Arzt trial in 2011 showing: (a) favorable position of the fetus while introducing the needle into the left ventricle, (b) catheter with balloon in left outflow tract and (c) dilatation of aortic valve by balloon insufflation.*[15]

remains. What if HLHS doesn't have to develop at all? It has been observed that anatomic cardiac obstructions that lead to ventricular dysfunction by diverting fetal blood flow in utero can result in cardiac chamber hypoplasia. Therefore, severe aortic stenosis (AS) in midgestation may lead to myocardial damage[8]. The abnormal flow dynamics and shear stresses appear to result in poorly orchestrated ventricular growth and development. As gestation continues, the cardiomyocytes undergo a switch in myogenic potential that lose the ability to undergo mitosis, meaning ventricular hyperplasia can no longer occur. Consequently, remodeling of the ventricular tissue is confined to muscular hypertrophy[6]. Therefore, due to faulty flow dynamics during development, a fetus progressively develops HLHS. This is possibly oversimplifying the pathogenesis as another likely cause could be a primary genetic disorder that affects the myocardial and valvular development[6]. There is some evidence of a link between HLHS and bicuspid aortic valve disease, as bicuspid aortic valves are the most common cardiac abnormality in first-degree relatives of children with HLHS[13]. Reports of bicuspid aortic valves in otherwise normal first-degree relatives to those with HLHS is as high as 11%[14], whereas bicuspid valves in the overall population is only 1-2%[6].

Thus it is theorised that if there can be early relief of AS then left-heart function may be preserved and HLHS could potentially be prevented[8]. This theory has been tested in clinical trials using FCI and has made tentative progress. One of the first larger clinical trials was done in 2004 by Tworetzky et al. in Boston. They performed balloon dilation in fetuses with severe AS in order to prevent HLHS. Twenty fetuses between 21 and 29 weeks gestation underwent the procedure. After both the mother and fetus were anaesthetized, a cannula was manually passed through the maternal abdomen and uterine wall and into the fetal chest under ultrasound guidance. The cannula then punctured the fetal LV and a coronary balloon was dilated while in the aortic valve (aortic valvuloplasty). Out of the 20 cases, 14 were considered technically successful; however, two of those babies died *in utero*. Of the 12 that survived, three went on to develop biventricular circulation at birth[8], meaning the LV was able to support systemic circulation, which is the optimal outcome[15].

Data collected from 2000 to 2009 demonstrated that 70 fetuses underwent the above procedure; 74% were considered to be a technical success, 29% of those were able to achieve biventricular circulation at birth[16] and another 8% were converted to a biventricular circulation after initial univentricular palliation[3].

In 2011, a trial done by Arzt et al. demonstrated that aortic valvuloplasty achieved technical success in 70% of the 24 procedures and biventricular circulation in 67% of those fetuses[15]. At a median follow-up of 27 months, 40% of those newborns had only AV balloon dilation in the first postnatal week and no further surgery. The remaining 60% had to undergo a Ross-Konno procedure, which is a

Table 2. Current Selection Guidelines for Fetal Aortic Valvuloplasty

Unequivocal AS (vs aortic atresia)
LV long axis Z score >−2
Threshold score ≥4 of the following
LV long axis Z score >0
LV short axis Z score >0
Aortic annulus Z score > −3.5
MV annulus Z score > −2
MR or AS maximum systolic gradient ≥20 mmHg

McElhinney et al.[16]

pulmonary valve autograft that is used to replace a diseased aortic valve[17].

Overall, these clinical trials have demonstrated not only improved outcomes throughout the years, but also that HLHS can indeed be prevented with the correct intervention. Makikallio et al. demonstrated that after successful FCI, the physiological parameters that lead to the development of HLHS were improved[18], indicating that progression to HLHS could be avoided. These parameters included improved aortic and mitral valve growth relative to control fetuses, as well as a clear increase in LV ejection fraction, anterograde flow in the transverse arch, and bidirectional flow across the foramen ovale[19]. However, there was no difference in growth velocity of the LV short or long axis[3]. Even if biventricular circulation is not achieved after FCI for AS, there is still improvement by the left side of the heart in contributing to the univentricular circulation, improving the heart's efficiency and durability[3].

Fetal Cardiac Intervention for HLHS with IAS

An area that needs special attention is a subset of infants who have HLHS, as well as a highly restrictive intact atrial septum (IAS). These neonates develop profound cyanosis and pulmonary oedema immediately after birth and resuscitative measures are often unsuccessful. Those who undergo emergency Norwood procedures have a mortality rate of 83% at 6 months[20]. Even neonates who underwent early transcatheter procedures to relieve atrial septal obstruction had a neonatal mortality of 48%[20].

The neonates who have HLHS with IAS suffer from high mortality rates due to intrapulmonary anatomic abnormalities, including "arterialization" of the pulmonary veins and lymphatic dilation due to left atrial (LA) hypertension *in utero*, subsequently suffering from severe cyanosis and pulmonary oedema[20]. It is thought that if prenatal LA hypertension causes these changes, the best form of management would be a procedure that creates an atrial septal defect *in utero*[20]. However, there are marked technical limitations to creating a large atrial communication. Recently, FCI was reported in 21 HLHS fetuses with IAS where the interatrial communication was created with either balloon dilation or placement of a stent. In two of the cases, the fetus died due to significant hemopericardium and those that were delivered had a surgical survival of only 58%[21]. However, there does seem to be some benefit to this procedure in terms of preoperative management, as these neonates with an atrial septal defect ≥3mm after FCI had higher oxygen saturation at birth and were less likely to need urgent postnatal left atrial decompression[22]. FCI may indeed be a useful procedure for this cohort of patients too, but more research is needed to refute or confirm this assumption.

Complications of FCI

FCI for aortic valvuloplasty is not without its complications. Almost half of all fetuses experience a combination of bradycardia and right ventricular dysfunction of variable severity[3]. Other complications include pericardial effusions, ventricular thrombosis and fetal death[23]. However, most of these complications are manageable. The bradycardia is treated with epinephrine administration into the left ventricle[23] or prophylactic administration of epinephrine and bicarbonate through the balloon catheter at the time of the intervention[3]. The thrombosis does not usually have any further consequences[23].

Furthermore, a concerted postnatal evaluation to determine if there are adverse neurological consequences of FCI has yet to be completed. However, a recent study illustrated that there is no evidence to suggest that prenatal aortic valvuloplasty significantly affects cerebral arterial flow parameters[24]. It should also be noted that no maternal complications requiring treatment have arisen during these trials[3,15].

Future considerations to improve FCI

One of the most important prerequisites for success is implementing the proper inclusion criteria to increase the chances of having an optimal biventricular outcome. The new inclusion criteria can be seen in Table 2. It was conceded that the loose entry criteria in the Boston 2004 study, including patients with heterogeneous cardiac anatomy[8], may have contributed to the poor outcomes reported for the procedure.

Gestational age at the time of the procedure should also be considered. The Arzt et al. group in 2011 had better success than the group in Boston in terms of optimal outcome since their cohorts had a higher gestational age and therefore a larger LV at the outset. A key factor in predicting success is the size of the LV at the time of the intervention[23]. Ultimately, safety and success rates are dependent on patient selection and the level of experience of the interventional team[15].

Other limitations in FCI include instrumentation and imaging techniques. Currently, with the exception of the 18-gauge curved tip cannula that was developed specifically for FCI, the instruments that are used are used off-label, which may hamper procedural feasibility and limit technical options[3]. As for imaging, ultrasound is used to manually guide the needle into the maternal abdomen through the chest cavity of the fetus and into the LV[15]. This poses a major barrier to technical success. It is a considerable challenge to accurately and rapidly deliver a needle across multiple tissue planes without damaging vital structures[25].

Emery et al. have begun to improve upon this technical limitation by creating a computer-assisted navigation (CANav) system that allows for the targeting of small structures during robotic surgery with minimally invasive interventional procedures. CANav system provides the user with an ultrasound image of the target structures but with the additional feature of visualizing the trajectory of the medical instrument before it is introduced into the body. That way, the needle's point of entry and trajectory can be adjusted prior to entering the body cavity of both the mother and the fetus. It also allows the operator confidence that the needle trajectory will target the desired structure and avoid any vital structures that could cause fetal complications[25].

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Measurement and Assessment of Swallowing: A Review

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> **"Swallowing is a complex, highly organised sequence of events that originates in the swallowing centre of the brain. …There has been a recent surge in the development of novel investigations exploring the swallow cycle."**

Abstract

Several different methods are available for examining the swallowing cycle and assessing dysfunction. The current gold standard and most widely used method is the modified barium swallow study, which uses videofluoroscopy to assess bolus transit through mouth, pharynx and upper oesophagus. Other methods include fibre-optic endoscopic evaluation, which uses a flexible endoscopy to observe the swallowing process. This method allows for precise visualisation of the anatomy. Surface electromyography and cervical auscultation, although not very specific, can be used as rapid screening tools for dysphagia. Manometry uses pressure sensors to record peristaltic activity in the pharynx and upper oesophagus and can be used in conjunction with video fluoroscopy (manufluorography) for more detailed information. The aim of this review article is to look at current methods for evaluating swallowing and to outline the advantages and disadvantages of each of these methods.

Introduction

Swallowing is a complex, highly organised sequence of events that originates in the swallowing centre of the brain. It is entirely under reflex control and is initiated by voluntary movement of a food bolus to the back of the throat. This causes sensory impulses to be generated that are subsequently transmitted to the medulla and lower pons, from which motor impulses then travel to pharyngeal and oesophageal muscles to initiate swallowing. During swallowing, respiration is reflexively inhibited to prevent food from being aspirated[4]. Problems with this complex process can lead to significant health issues including aspiration pneumonia, dehydration, malnutrition and reduction in quality of life. Difficulty in swallowing is termed "dysphagia".

Definition

Dysphagia is defined as an abnormality in the transfer of a bolus from the mouth to the stomach[5]. It is associated with a sensation that solids or liquids are not being swallowed correctly.

Aetiology and Classification

Dysphagia is not a single disease entity, but a clinical manifestation of any problem involving the complex swallowing mechanism[6]. It is therefore important to determine the underlying causes.

1. Neurogenic dysphagia

Stroke is the most common cause of neurogenic dysphagia, with 30–40% of stroke victims suffering from significant dysphagia. As many as 20% will die in the first year from aspiration pneumonia secondary to the dysphagia[7]. Parkinson's disease causes degeneration of subcortical neurons (especially in the substantia nigra), leading to progressive motor deficits. In later stages it can lead to dysphagia due to dysfunction of oral, pharyngeal and oesophageal muscles[8]. Lower motor neuron deficits such as myasthenia gravis, Eaton-Lambert syndrome, amyotrophic lateral sclerosis and multiple sclerosis may equally lead to significant dysphagia if they affect neurons supplying oral, pharyngeal or oesophageal musculature; nerves in the swallowing centre may also be implicated. Traumatic brain injury is another major cause of neurogenic dysphagia. Finally, iatrogenic or congenital recurrent laryngeal nerve paralysis may also cause transient or permanent dysphagia. Unilateral paralysis is usually self-limiting and less severe in nature.

2. Structural/Mechanical dysphagia

The most common cause of mechanical swallowing difficulty is surgical resection of head and neck cancers. Tumours of this area include squamous cell carcinoma, thyroid carcinoma, adenocarcinoma and neuroendocrine neoplasia. Removal of parts of the tongue (glossectomy), larynx (laryngectomy) or even the oesophagus (oesophagectomy) may cause problems with swallowing. Other mechanical causes for dysphagia include infection/inflammation such as tonsillitis, epiglottitis, pharyngitis or quinsy (this may cause trismus – an inability to fully open one's mouth).

3. Oesophageal dysphagia

Oesophageal dysphagia refers to any cause of dysphagia that originates in the oesophagus. This may occur when the lumen becomes stenotic after swallowing a bolus that is too large for the oesophageal lumen. Another common cause of this is the formation of rings and webs of abnormally thick mucosal bands, causing narrowing in patients with iron deficiency anaemia. Furthermore, strictures may occur due to irradiation in patients being treated for head and neck cancers, or by acid reflux into the oesophagus, which is known as gastro-oesophageal reflux disease (GORD). Finally, sphincteric problems might occur at the lower end of the oesophagus, which can cause late regurgitation of food. This is called achalasia and is due to a non-relaxing lower oesophageal sphincter. A barium swallow test shows a classic dilated oesophagus with "parrot-beaked" tapering at the junction.

Swallowing Measurement

Several different methods for evaluating swallow and dysphagia have been described. Preceding any investigation is a thorough history and examination by ear, nose and throat specialists, speech and language therapists and nurses. Some studies offer subjective information that requires further evaluation (e.g. modified barium swallow study) and others determine quantitative values (e.g. impedance pharyngography[9] and studies to measure average volume per swallow, speed per swallow and swallowing capacity[10]). Clinical examination is able to identify only 60% of people who aspirate, leaving 40% undiagnosed and vulnerable to significant complications[11]. Thus, the need for effective diagnostic tools is great.

1. Modified Barium Swallow (MBS) Study

This investigation involves the use of video fluoroscopy to examine anatomical or physiological deficits along the oropharynx and monitor

improvements in rehabilitation[12]. This procedure is widely available and can be used in patients of all ages. It is considered the gold standard for evaluating not only swallow dysfunction, but the mechanism of swallow itself[6]. The examination is done in the upright position and examined laterally as well as anteroposteriorly. The patient is given a bolus of thin liquid to swallow orally. This liquid contains barium and represents saliva. The patient first takes 1mL and then progresses to 2mL, 5mL and finally 10mL. Should the patient fail at one stage of the test, measures such as postural techniques or sensory enhancement are taken to help them to swallow the bolus[12]. The examination is done by videofluoroscopy, which films the entire swallowing sequence. The average swallowing cycle requires only 1.0–1.5sec (with the oral phase only lasting about 0.5sec). This requires a high frame rate that only filming can provide (30 frames/s). Using this method the cycle can be carefully examined frame-by-frame (refer to Figure 1)[13].

Videofluoroscopy also provides information on transit time, motility problems and amount and aetiology of aspiration. The patient only receives only small amounts of radiation during the procedure[14].

A study of 608 patients by Martin-Harris et al. aimed to examine the clinical utility and yield of the MBS study[15]. They found some degree of abnormality in 90% of patients examined. Aspiration occurred in 32% and abnormal swallowing physiology was identified in 57% of those in which aspiration did not occur. Appropriate referral to other specialties (e.g. gastroenterology or speech and language therapy) after examination occurred in most cases. They concluded that MBS has a low false-positive rate and is a cost-effective investigation when compared to bedside observations or medical diagnosis[15].

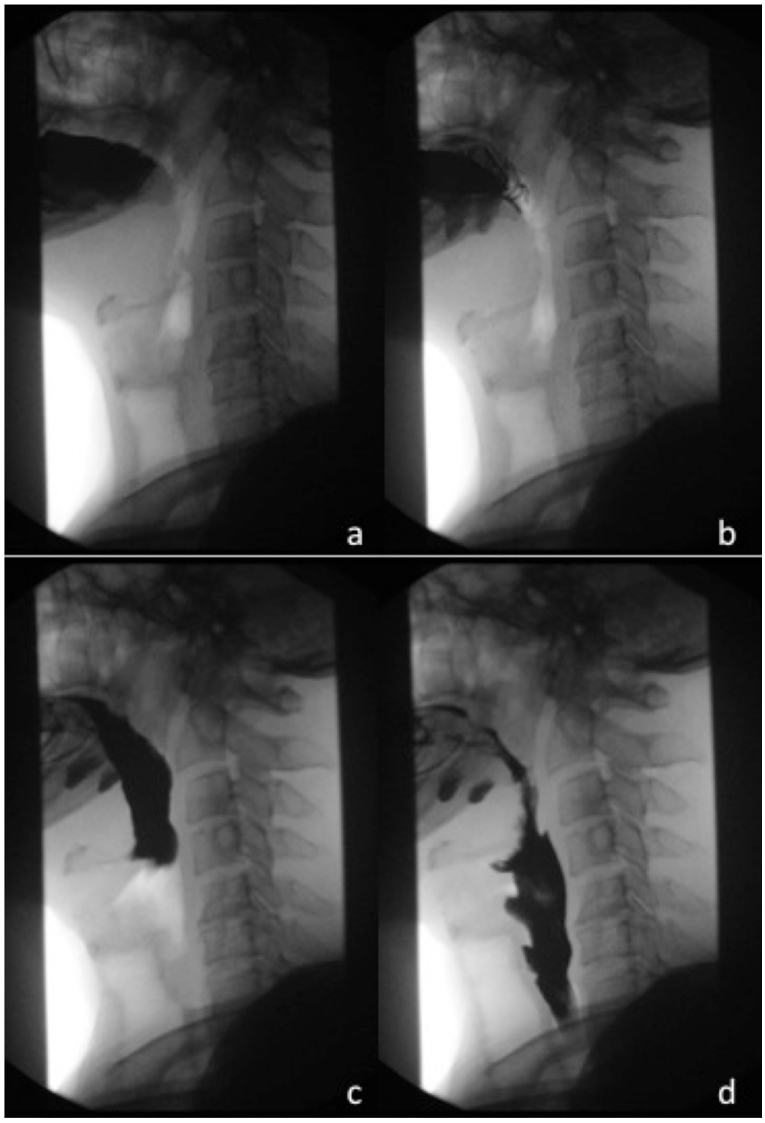

Figure 1. *Lateral radiographic overview of a normal swallowing sequence of a healthy patient with 10mL of barium. (a) Barium bolus is positioned in the mouth and is sealed off from the throat by approximation of the tongue with the soft palate. (b) The soft palate opens, allowing barium into the oropharynx. (c) Pharyngeal phase of swallowing has commenced. The upper oesophageal sphincter is open allowing free passage of fluid. (d) Peristaltic activity in the cervical oesophagus propels bolus into stomach. Adapted from Hellerhoff*[1]

2. Fibre-Optic Endoscopic Evaluation of Swallowing (FEES)

This assessment was first described by Langmore, Schatz and Olsen in 1988 and has become increasingly popular for assessing oral cavity and pharyngeal anatomy[16]. It was developed to evaluate the swallow mechanism if the more expensive videofluoroscopy was not available.

This procedure uses a flexible fibre-optic pharyngolaryngoscope (3.7mm diameter) to gain access to the pharyngeal space. No anaesthetic is required. The patient is seated upright (or at

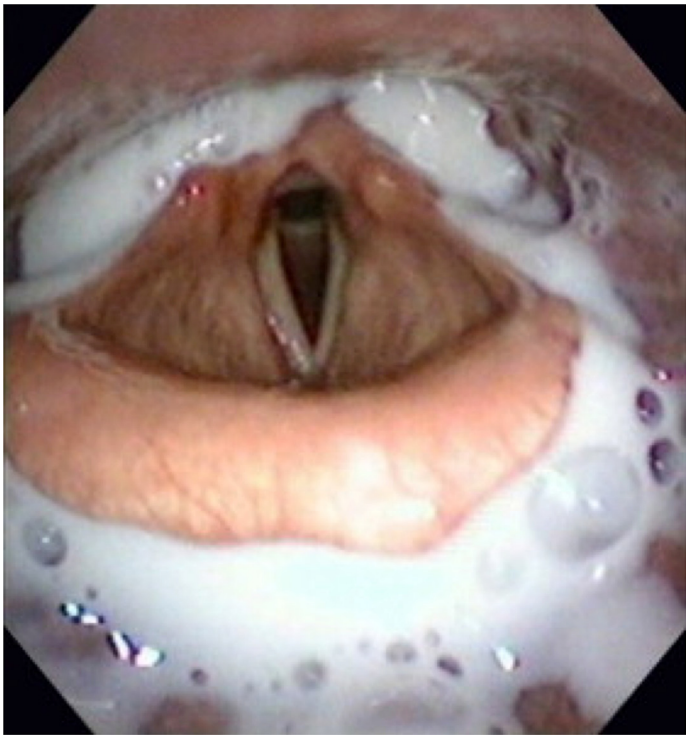

Figure 2. FEES image of the vocal folds. White liquid spillage into vallecula and pyriform fossae. Epiglottis anteriorly (bottom of picture). Adapted from Leder, S.B. and Murray, J. T.[3]

45° if bed-bound) and the scope is introduced into the nares. The scope is advanced below the inferior turbinate into the nasopharynx, where the velopharyngeal competence can be assessed. Moving through the oropharynx, the scope is finally passed to a point posterior to the epiglottis, giving clear view of the vocal cords.

With the scope in this position, the swallow assessment can commence: measured quantities of food/liquid (5-10ml) are given to the patient to swallow. The substances are dyed with green or blue food colouring. The bolus cannot be monitored during pharyngeal swallow as the substance obstructs the view of the scope. However, the phases immediately preceding and following swallow can be observed and provide useful insight into premature spillage and residual food in the pharynx[16,14,17].

FEES is more sensitive than MBS in examining anatomy and can identify subtle anomalies. It provides limited information on the oral phase of swallowing and is unable to examine the pharyngeal phase (Figure 2)[17]. There is no radiation exposure and it can be performed faster and with less preparation than videofluoroscopy, making it ideal when immediate information is required.

A study by Kelly et al. found that, in general, pharyngeal residue was perceived to be greater in FEES than in videofluoroscopic studies[18]. Their study concluded that FEES and videofluoroscopy cannot be used interchangeably, but should be used to complement each other. An advantage of FEES is its use in patients who are bed-bound and cannot sit up for a barium swallow test.

3. Scintigraphy

Scintigraphy is a nuclear medicine test used to track the movements of a bolus and effectively quantify residual volumes in the oropharynx, pharynx, larynx, trachea and/or lungs. During the test, the patient swallows a bolus of radioactive liquid (containing technetium 99m) and this is recorded by a gamma camera. For the swallow phase, data acquisition is continuous (25 frames/s) and usually involves the oral cavity, thoracic cavity and, less frequently, the upper abdominal cavity. Static images at 15-30 min intervals are acquired for several hours after the test. Results are reported on time-activity curves[19]. This yields a quantitative image of transit and metabolic processes, and the amount of aspiration and residue can be measured[17]. This investigation does not provide information on the anatomy and physiology of the underlying mechanism for dysphagia. It is particularly useful for the assessment of GORD. If lungs and airways are clear of material immediately after swallowing, but it is accumulating over time, GORD is the cause of the aspiration[19].

4. Ultrasound

Ultrasound has been used to observe tongue function, motion of the hyoid bone, and to measure oral transit times. It has limited capacity for assessing the pharynx due to the different tissue types that are located in the neck as well as the deep location of the pharynx. Ultrasound remains useful for assessing tongue movement during the preparatory and oral phase of swallowing[20].

5. Electromyography

Surface electromyography (SEMG) can be used to measure electrical surface activity of the muscles involved in swallowing. This investigation relies on the presumption that different diseases and patterns of dysphagia have unique SEMG patterns[21]. Surface electrodes are placed on the skin above the muscle being investigated. Floor-of-mouth or laryngeal elevation muscles have been used most frequently for this test, as they are closest to the surface. They provide good information about the initiation and oral phase of swallowing. SEMG cannot detect activity in pharyngeal musculature as the muscles are situated too deeply.

An alternative electromyographic method uses hooked-wire electrodes to assess superior pharyngeal constrictor muscle activity during swallowing[22]. This method is very invasive and, given that less invasive tests with better results are available, this test is useful only for very specific cases where dysfunction of specific swallow muscles must be assessed. SEMG, although not very useful for diagnosis, can provide a quick and easy screening test with high sensitivity but low specificity for dysphagia or odynophagia[21].

6. Swallowing Sounds (Acoustics)

The goal of cervical auscultation is to establish physical parameters of swallowing sounds that are characteristic of dysphagia. These parameters can then be evaluated objectively via acoustic analysis. The advantages of such a system is that it is non-invasive, inexpensive, objective, can be performed at the bedside and does not expose the patient to radiation[23].

This technique uses microphones placed on the neck to record the swallowing sound. The most widely accepted position with the least amount of noise (generated by the carotid artery and laryngeal elevation) is the lateral border of the trachea, just inferior to the cricoid cartilage[23,24]. The patients are asked to swallow different-sized boluses in one complete action. In a study by Santamato et al., every swallow was recorded and three parameters were established as the acoustic profile: firstly, the duration of the swallowing sound (DSS) measured in miliseconds (ms) and defined as the time between start and the end of the acoustic signal; secondly, the peak intensity (PI) measured in decibels (dB) and defined as the highest displacement of the acoustic signal; and finally, the peak frequency (PF) measured in Hertz (Hz). The DSS in patients with neurogenic dysphagia was significantly increased when compared to healthy patients (1402 msec and 440 msec respectively; $p<0.01$)[24].

Although this test manages to objectively quantify certain parameters, it is still difficult to correlate an acoustic signal to an anatomical event. The efficacy of this test is also affected by inter-patient variation of the previously mentioned parameters, as well as by age[25].

7. Manometry

Oesophageal manometry is routinely used to establish oesophageal function. It uses a pressure sensor and transducer to relay pressure information within the oesophagus. This pressure represents peristaltic function and sphincteric competence[26]. It is a useful tool in the diagnosis of GORD, achalasia and dysphagia which is caused by dysfunction of the upper oesophageal sphincter. It cannot, however, monitor the entire swallowing process.

Pharyngeal manometry records pressure changes in the pharynx during the swallowing cycle. It requires solid-state pressure sensors with a fast frequency response. Unlike oesophageal manometry, where transit time ranges from 8-20s, pharyngeal transit time is significantly shorter at 0.5-1.5s. Three pressure sensors are placed transnasally: one at the base of the tongue, another at the upper oesophageal sphincter and a third at the upper oesophagus. This investigation is often done concurrently with video fluoroscopy to correlate pressure change with anatomical and physiological processes and events.

Until recently, pharyngeal manometry was not widely used as the resolution of the solid-state

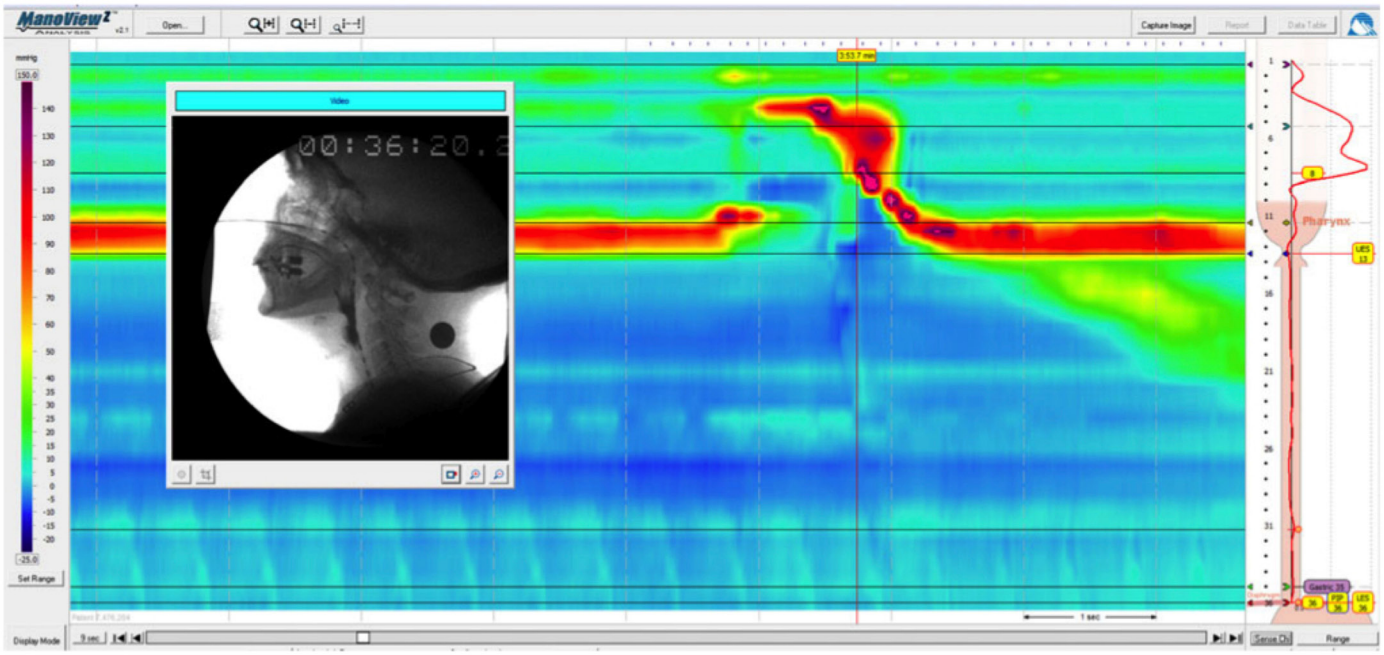

Figure 3. *Screenshot from ManoView™. X-axis: time, Y-axis: sensor position (distance). Moving the red line advances the video-fluorography and the topographical map. The numbers on the right denote the sensor number. Adapted from Nativ-Zeltzer et al.*[2]

pressure sensors was very low. In addition, it was difficult to correlate pressure changes with anatomical events. High-resolution manometry uses more sensors (30-36 compared to 3-5 in older devices) that are smaller and placed much more closely together (<1cm apart) than those of older manometric systems[2]. The upper oesophageal sphincter can move up to 3cm during swallowing[27], causing it to move in relation to the old sensors and possibly even miss them. The new system resolves the issue of having to place sensors in an exact location to yield useful data.

Along with the improvement of manometry, new sensory software has been produced which allows concurrent videofluorographic and manometric observation of the swallowing process (ManoView™).

The two measurement modalities together provide a more in-depth view into the process of swallowing and its dysfunction in dysphagia. Manometry alone cannot observe the oral phase of swallow, cannot determine if there is any residue present, and lacks mechanical accuracy. It does aid videofluoroscopy in objectively quantifying pressure vectors that are affecting the bolus as well as provide subtle cues as to the nature of the dysphagia[2].

Conclusion

There has been a recent surge in the development of novel investigations exploring the swallow cycle. MBS is a relatively expensive procedure, requiring elaborate equipment, several experts from different fields and a precise experimental regime. So, although it is the current gold standard for diagnosing dysphagia, there is a demand for effective alternative methods offering faster and cheaper screening of pharyngeal function. MBS is not available in every hospital/clinic and, in some situations (e.g. bed-bound patient), cannot be used at all. Manometry is the most sophisticated investigation and yields most data for one swallow cycle. It may be used to diagnose more complex disease or pharyngeal dysfunctions.

In summary, no single procedure is perfect for examining swallow. Rather than looking at each procedure as an exclusive diagnostic tool, they should be viewed as adjuvants to each other in the diagnosis of complex swallowing disorders.

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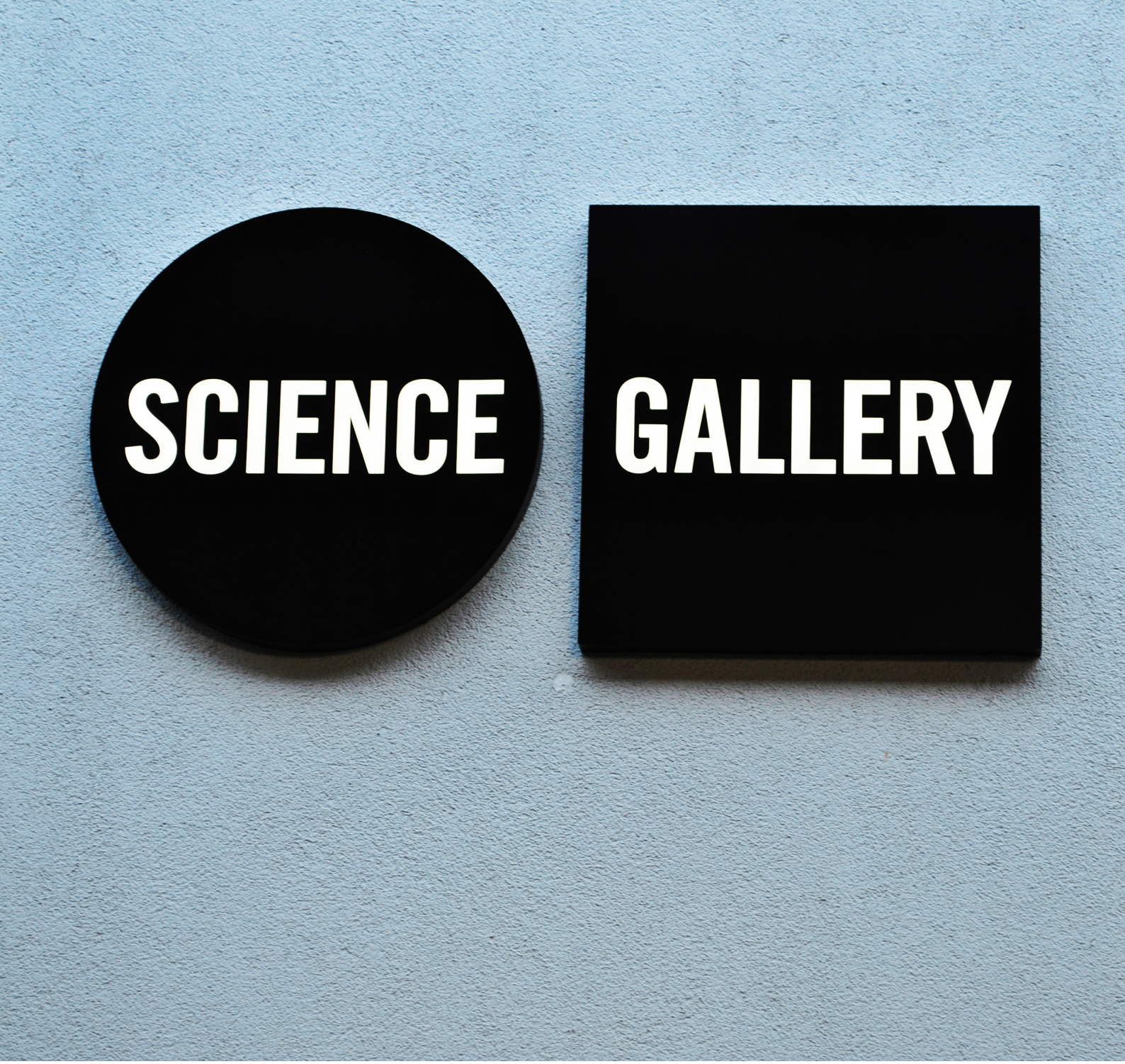

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