



Chiari
Malformation
Syringomyelia

SYRINGOMYELIA-CHIARI 2018 INTERNATIONAL SYMPOSIUM

TUESDAY 17TH – FRIDAY 20TH JULY 2018

MACDONALD BURLINGTON HOTEL, BIRMINGHAM, ENGLAND

WELCOME

A very warm welcome to Birmingham and to Syringomyelia-Chiari 2018. Many of you have travelled long distances in order to attend this meeting and for this we offer our sincere thanks. As a patient myself I can only express my admiration for and thanks to those of you who take an interest in these conditions and who try to help those of us living with the effects of Chiari and syringomyelia.

For many years the Ann Conroy Trust (formerly known as Ann's Neurological Trust Society) has provided support for patients with these conditions. It also engages in various educational programmes, aimed at patients and their families and at healthcare professionals. Some of these are local, UK-based events but others, such as this meeting, are aimed at the international medical community, intending to foster free exchanges of ideas and experiences for the benefit of all patients.

This meeting has been co-organised by the Ann Conroy Trust and Aesculap Academia. We have tried to put together an event that will provide you with edification and professional stimulation, as well as some recreation. I very much hope that you will enjoy your stay with us.



Lynn Burton
Chairman
Ann Conroy Trust

INTRODUCTION

Syringomyelia-Chiari 2018 has its roots in the very successful symposium Syringomyelia 2007, held in Rugby, England. This meeting was followed by Syringomyelia 2010, held in Berlin and then Syringomyelia 2013, held in Sydney. A further event was held in Long Island, New York, in 2017, organised by the American Syringomyelia-Chiari Alliance Project. This conference benefitted from a series of platform speeches delivered by invited international experts. Particular attention was given to areas of controversy, including the relationship between Chiari malformations and various other medical conditions. Another feature of the Long Island meeting was the participation by a number of patients, many of whom were affected by one or more of these conditions.

Syringomyelia-Chiari will also look at some of these areas of controversy and it will also have the benefit of patient representation from a number of national support groups. In addition, it aims to provide a forum for the presentation of free papers, in the manner of the earlier conferences. An awareness of what other, "jobbing" clinicians are doing and achieving, in different parts of the world, is always likely to be of benefit to any specialist who is called upon to treat these enigmatic conditions. Such benefits can, hopefully, then be incorporated into daily patient care.

In naming the present conference Syringomyelia-Chiari 2018 we are acknowledging that many more cases of hindbrain hernias without an associated syrinx cavity are now being detected. This is thanks to the widespread and ready availability of MR imaging. As a result, Chiari is now seen as a condition in its own right, not just the commonest cause of syringomyelia. The filling mechanism underlying syringomyelia formation may remain a source of fascination for neuroscientists and clinicians but hindbrain hernias, in their various forms, have become the source of distress for a good many patients.

The venue for Syringomyelia-Chiari 2018 was chosen because of its central location in the City of Birmingham, immediately opposite the main railway station and in close proximity to shops, bars and restaurants, as well as museums and galleries. We hope that you will have time to avail yourself of some of these facilities and that, together with what you learn at the symposium, you will be left with good memories of Syringomyelia-Chiari 2018.



Graham Flint
Consultant Neurosurgeon
Birmingham, England

INVITED SPEAKERS

Fiona Adams, Birmingham, England

Paolo Bolognese, New York, USA

Andrew Brodbelt, Liverpool, England

Palma Ciaramitaro, Turin, Italy

James van Dellen, London, England

Atul Goel, Mumbai, India

John Heiss, Bethesda, USA

Nicholas Higgins, Cambridge, England

Jörg Klekamp, Quakenbrueck, Germany

Anita Krishnan, Liverpool, England

Shailendra Magdum, Oxford, England

Christopher Parks, Liverpool, England

Clare Rusbridge, Guildford, England

Guirish Solanki, Birmingham, England

Stavros Stivaros, Manchester, England

Marcus Stoodley, Sydney, Australia



- The first long term telemetric pressure sensor for the treatment of hydrocephalus
- Decision-making support with quick and easy evaluation of clinical symptoms
- Determination of optimal valve pressure setting
- Localisation of shunt blockages: Proximal and distal



MIETHKE SENSOR RESERVOIR

TELEMETRIC SHUNT CONTROL – READING INNER VALUES

MEMORIAL LECTURES

Amongst the invited lecturers who have given their time to address this conference we are privileged to have three individuals of standing in the field of syringomyelia and Chiari, who have each kindly agreed to deliver one of our three eponymous lectures. To accompany these lectures we provide the following eulogies:

EDWARD OLDFIELD 1947–2017

Edward H Oldfield, MD was a highly creative and productive neuroscientist and neurosurgeon, leading research programs that changed the modern surgical treatment of patients with pituitary tumours in Cushing's disease, with brain and spinal cord tumours in von Hippel-Lindau disease, and with spinal arteriovenous malformations.

Oldfield completed two years of surgical residency training at Vanderbilt University, before spending a year as a visiting registrar in Neurology and Neurosurgery at the National Hospital for Nervous Disease in Queen Square, London, England. He then completed neurosurgical residency training at Vanderbilt University. He spent a year in the private practice of medicine before coming to the National Institutes of Health in 1981. He spent most of his research career there, in the Surgical Neurology Branch, the National Institute of Neurological Diseases and Stroke. He started as a senior staff fellow, became chief of the clinical neurosurgery section in 1984, and served as chief of the surgical neurology branch from 1986 until 2007. He received the Public Health Superior Service Award in 1991, for successfully managing the surgical neurology branch, training academic neurosurgeons and advancing the understanding of the biology of brain tumours. He retired from full-time government service in 2007 to become the Crutchfield Chair in Neurosurgery and Professor of Neurosurgery and Internal Medicine at the University of Virginia.

Among his many research accomplishments, Oldfield's research led to new insights into how Chiari I malformation causes syringomyelia. He developed a new drug-delivery technique, called convection-enhanced delivery, for treatment of central nervous system diseases, including brain tumours, Parkinson's disease, and lysosomal storage diseases. His laboratory developed gene therapy for malignant brain tumours. He directed the first clinical trial of gene therapy within the central nervous system. He discovered that reduced nitric oxide around cerebral arteries produced cerebral vasospasm after aneurysmal subarachnoid haemorrhage. He followed-up this finding with clinical trials of novel agents treating cerebral vasospasm.

Oldfield was a highly acclaimed academic neurosurgeon, serving on the editorial boards of prominent neurosurgical journals, including as co-chairman of the Journal of Neurosurgery, from 2001 to 2002, and associate editor from 2009. He served as the vice-president and president of the Society of Neurological Surgeons and received the Grass Medal for Meritorious Research in Neurological Science, from that organization. The American Association of Neurological Surgeons awarded him the Farber Award for his brain tumour research, the Harvey Cushing Medal (its highest honour) for "his many years of outstanding leadership, dedication, and contributions to the field of neurosurgery" and the Cushing Award for Technical Excellence and Innovation in Neurosurgery. The University of Kentucky Medical Alumni Association recognized him as "the quintessential clinical-scientist" who made "remarkable contributions to the understanding of the nervous system and the practice of neurosurgery."

Dr Oldfield authored over 500 scientific and clinical articles and was co-inventor of patents on convection-enhanced drug delivery and genetic therapy. He fostered the career development of his fellows and other trainees, many of whom achieved tenured positions and chairmanships in neurosurgery departments in the United States and internationally.

Dr Oldfield is survived by his devoted wife, Susan (Wachs), and his loving daughter Caroline. His family welcomed colleagues and trainees to their home, creating life-long friendships that extended beyond science and medicine. His family, friends, professional colleagues, and patients will miss his kindness, advice, concern, and care.

JOHN HEISS

MEMORIAL LECTURES

BERNARD WILLIAMS

1932-1995

Bernard Williams was born in 1932, in a place called Stockport, not far from the city of Manchester, in the north of England. He studied medicine in Birmingham, qualifying in 1955. After military service he began his training in neurosurgery, initially at the National Hospital, London and then in Birmingham once more. His first consultant post was back up in the north of the country, in the city of Hull, lying on the Humber estuary. He worked there for three years, before returning to the English Midlands, to take up a post at the Midland Centre for Neurosurgery and Neurology, situated in the town of Smethwick, in the Black Country, adjacent to the Greater Birmingham conurbation. He continued to work there until his untimely death in 1995, at the age of 63.

Bernard first became interested in the condition of syringomyelia during his training. He subsequently carried out pioneering research into the condition, publishing seminal papers and establishing an international reputation in the field. He remained fascinated by the condition throughout his career. He is still quoted widely in neurosurgical literature. He also made important contributions to our understanding of other disorders of cerebrospinal fluid circulation, as well as other neurosurgical diseases. His work was recognised by the award of the Cassey-Holter memorial prize, in 1977 and the Pudenz prize in 1994. He received further honours from the Royal College of Surgeons of England.

Bernard Williams possessed a remarkable intelligence, despite which he remained a modest, often self-effacing person, with no hint of arrogance or pomposity. He encouraged students and young doctors to see the fascination in clinical neurological science and to share his enthusiasm for this field. He had an open-minded approach to whatever subject he considered and was always completely honest in what he said. When operating he paid meticulous attention to detail. When results were not as good he wanted he would agonise over the reasons and always seek to improve himself.

Outside the world of neurosurgery, Bernard Williams had a passion for the game of chess, a talent supported by his extraordinary memory. It was therefore very fitting that his widow chose a chess piece as the headstone for his grave. Bernard died riding his motorbike, a victim of the impetuous haste of morning rush-hour traffic. Bernard left behind him four children by his first marriage and two from his second. There are also many of his former patients who remember him with fondness and gratitude. So too do many neurosurgeons who trained under his direction. There are no lengthy, wordy inscriptions on his grave, just his dates, preceded by what he wrote at the bottom of any letters – "Bernard Williams, Neurosurgeon".

GRAHAM FLINT

ANN CONROY

1943-1992

Ann Conroy was born in 1943, in the city of Leicester, in the English Midlands. She developed scoliosis in her teens but, in the pre-MRI era, her syringomyelia was not diagnosed until she was in her early thirties. She underwent surgery for the underlying hindbrain hernia shortly afterwards. Although improved after the operation, Ann remained crippled but bore this burden with fortitude and without complaint, preferring instead to get on with life as best she could.

I did not have the privilege of knowing Ann but I recall Bernard Williams talking about her. He described a remarkable and energetic lady who, despite having very significant neurological disabilities, was determined to set-up an organisation that would help others who also suffered from syringomyelia. Ann, it would seem, was impressed by two things. One was the seeming lack of knowledge about or understanding of syringomyelia and Chiari amongst health care professionals, including neurologists and neurosurgeons. The other was Bernard Williams and the dedication that he applied in attempting to understand and treat syringomyelia. Crippled though she was, as a result of her condition, Ann set about founding the charity which now bears her name. Her aims were to fund Bernard's continuing work and to provide support for other people affected by these conditions.

From its foundation, as "Ann's Neurological Trust Society", the now re-named Ann Conroy Trust has developed, over the past 40 years, into a highly successful support organisation for people living with syringomyelia or Chiari. It has also organised teaching events for health-care professionals, both at a national and an international level. It supported the creation and publication, in 2014, of the monograph entitled "Syringomyelia: a disorder of CSF circulation". In this volume is a chapter entitled "Historical vignettes". The account therein of Ann Conroy's life was derived from a eulogy written about her, following her death in 1992, by Bernard Williams. His final sentence reads: "She may not ever have been able to work or to marry or to bear children and she may never have travelled far in her lifetime, but the journey of her spirit was immense. The work that she began will surely continue".

Continue it does, with Syringomyelia-Chiari 2018.

GRAHAM FLINT

INVITED LECTURERS

FIONA ADAMS



Fiona Adams is a specialist occupational therapist who works in the Regional Posture and Mobility Service, part of the Birmingham Community Health Care Trust. She specialises in the field of complex postural mobility provision and provision of manual mobile arm equipment. She works with all types of conditions and age groups. She is particularly passionate about the effects of fatigue on people and the importance of fatigue management in treating their conditions.

PAOLO BOLOGNESE



A native of Torino, Italy, Paolo Bolognese graduated in 1986 from the Medical School of the University of Turin. In 1990, he completed his first neurosurgical training at the same university under the guidance of Professor Victor Fasano. In 1992 he accepted the invitation of Dr Thomas Milhorat to transfer his on-going laser Doppler research to the United States, at the SUNY Health Science Center at Brooklyn. There he completed his second residency in neurosurgery and his fellowship in the management of Chiari I malformation and related disorders. In 2001 Dr. Bolognese joined Dr. Milhorat at North Shore University Hospital in Long Island, New York, where together they founded The Chiari Institute. Dr. Bolognese remained there until 2014, first with Dr. Milhorat and, then with Dr. Harold Rekate. In 2014 he started the Chiari Neurosurgical Center, where he was later joined by Dr Roger Kula. Dr Bolognese has performed 1,400 Chiari decompressions, 900 craniocervical fusions, and 900 cord de-tetherings. He is on the scientific educational advisory board of the Chiari Syringomyelia Foundation and on the board of directors of the American Chiari and Syringomyelia Alliance Project.

ANDREW BRODBELT



Andrew Brodbelt is a consultant neurosurgeon and Divisional Clinical Director at the Walton Centre NHS Foundation Trust in Liverpool UK, and an honorary clinical senior lecturer at the University of Liverpool. His neurosurgical training was in Liverpool UK, and his PhD 'Investigations in post-traumatic syringomyelia' was completed in Sydney, Australia with Prof Marcus Stoodley. He has published 13 papers and 3 book chapters on Chiari and syringomyelia. His research interests are in the pathophysiology of syringomyelia, the underlying biomechanics, and treatment outcomes. He is married to Kathryn, and has three children, Edward, Jessica, and Harrison. He enjoys skiing, mountain biking, road cycling, mountain walking, and triathlons.

PALMA CIARAMITARO



Palma Ciaramitaro is a neurologist working as a consultant at the Expertise Center for Syringomyelia and Chiari Syndrome-CRESSC, in the Neuroscience Department of the University of Turin. She is chairman of the Rare Disease Committee for the Turin Medical College and coordinator of the Italian Chiari and Syringomyelia Consortium. She is a member of the Italian Neurological Society, the Italian Clinical Neurophysiology Society, the Italian Neuropsychology Society and the Peripheral Nerve Society. Palma has 90 publications, including 50 papers on Journals listed on Current Contents, as well as chapters on neurological books. Her main fields of interest are clinical neurophysiology, rare neurological diseases, neuropathic pain, movement disorders and epilepsy.

INVITED LECTURERS

JAMES VAN DELLEN



James van Dellen has practiced neurosurgery in South Africa, the USA and the UK. He knew Bernard Williams and has been involved with the Ann Conroy Trust since its emergence in its current form and activities. He has for some years explored and followed the manner in which Chiari-syringomyelia might change and impact on family planning, pregnancy and parturition, as a sub-interest in the subject.

ATUL GOEL



Atul Goel is Professor and Head of Department at King Edward Memorial Hospital and Seth G.S. Medical College in Mumbai, India. He is consultant neurosurgeon at the TATA Memorial Hospital and Cancer Research Institute. He has held posts on the editorial boards of numerous neurosurgical journals, variously as chairman, editor or advisory board member. He has published two specialist neurosurgical textbooks and approaching 600 papers in medical journals. He has been twice a recipient of the Indian Council of Medical Research National Award, for outstanding scientific research and for outstanding research in neurology.

JOHN HEISS



John Heiss is the Chair of the surgical neurology branch and director of the neurological surgery residency training program in the National Institute of Neurological Disorders and Stroke at the National Institutes of Health, Bethesda, Maryland, USA. He received BS and MD degrees from the University of Michigan and completed neurosurgery residency training at the University of Cincinnati College of Medicine. He is vice-chair of the Combined Neurosciences Institutional Review Board at the National Institutes of Health. He is board certified in neurological surgery. He is principal investigator of studies into the natural history and genetics of Chiari I malformation and syringomyelia.

JÖRG KLEKAMP



Jörg Klekamp studied medicine at the Medical School of Hannover, Germany. Between 1986 and 1988 he carried out clinical and experimental work in the department of neuropathology at Sydney University, Australia. Between 1989 and 2003 he carried out clinical work in the department of neurosurgery at Nordstadt Hospital, Hannover, under the supervision of Professor Samii. In 1995 he carried out experimental and clinical studies on syringomyelia at the University of California in Los Angeles, working with Professor Batzdorf. In 1995 he was presented with the Wilhelm-Tönnis Award by the German Society of Neurosurgery, for clinical and experimental studies on syringomyelia. In 1997 he worked in the department of paediatric neurosurgery at Beth Israel Hospital in New York, under the supervision of Professor Epstein. Since 2002 he has been associate professor at the Hannover Medical School and, since 2004, neurosurgeon at Christliches Krankenhaus, Quakenbrück.

INVITED LECTURERS

ANITA KRISHNAN



Anita Krishnan is a consultant neurologist at the Walton Centre NHS Foundation Trust, Liverpool, England. Her clinical interests are in the fields of headache medicine and cerebrospinal fluid pathway disorders, especially idiopathic intracranial hypertension.

Along with running general neurology clinics and managing neurology in-patients, she also conducts tertiary specialist clinics in complex headache disorders. She has drawn up the regional secondary care pathways for management of acute headache and an approach to papilloedema. She teaches extensively all cadres of medical professionals on the recognition and management of primary and secondary headache. She is a member of the Association of British Neurologists advisory group on headaches and the special interest group in idiopathic intracranial hypertension and is one of the contributing authors to the international consensus guidelines on this condition.

SHAILENDRA MAGDUM



Shailendra Magdum trained in Mumbai, Hull, Sheffield and Oxford. He has been a consultant neurosurgeon since 2006, originally at Birmingham Children's Hospital before moving to Oxford in 2009, where he is currently employed as a consultant paediatric neurosurgeon to the Oxford Radcliffe Hospitals NHS Trust. His practice is predominantly paediatric and craniofacial neurosurgery. He has particular expertise in surgery for craniosynostosis, craniofacial malformations and Chiari. He views the Chiari 1 complex in children as presenting in a different way, depending on the age of the patient. Surgery for Chiari 1 in early years has its own unique natural history which needs to be studied in detail.

CHRISTOPHER PARKS



Chris Parks trained at Imperial College School of Medicine in London. He went on to basic surgical training in the Wessex region and then specialist training in Manchester and the North West of England. He completed his paediatric neurosurgical fellowship in Alder Hey children's hospital, in Liverpool, where he now works as a consultant paediatric neurosurgeon. Chris has also been on visiting fellowships in Uganda, Rotterdam and Memphis. Chris is craniofacial lead and trauma lead for neurosurgery in Alder Hey. He has a busy general neurosurgery practice but also has a special interest in complex cervical spine surgery, cranio-cervical junction anomalies, neuro-endoscopy and neuro-oncology.

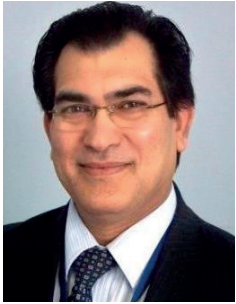
CLARE RUSBRIDGE



Clare Rusbridge graduated from the University of Glasgow in 1991 and, following an internship at the University of Pennsylvania and general practice in Cambridgeshire, she completed a BSAVA/ Petsavers residency and was staff clinician in neurology at the Royal Veterinary College. She became a Diplomat of the European College of Veterinary Neurology in 1996 and a RCVS Specialist in 1999. In 2007 she was awarded a PhD from Utrecht University for her thesis on Chiari-like malformation & syringomyelia. For 16 years she operated a neurology and neurosurgery referral service at the Stone Lion Veterinary Hospital in Wimbledon. In September 2013 Clare joined Fitzpatrick Referrals and the University of Surrey. Her professional interests include epilepsy, neuropathic pain, inherited diseases, and rehabilitation following spinal injury. She was awarded the J. A. Wright (a.k.a. James Herriot) Memorial Award by The Blue Cross animal welfare charity in 2011 for her work with syringomyelia. Clare has authored or co-authored over 90 scientific articles and book chapters, including being co-editor for a medical textbook on Syringomyelia, published by Springer in 2014.

INVITED LECTURERS

GUIRISH SOLANKI



Guirish Solanki is a consultant paediatric neurosurgeon at the Birmingham Children's Hospital, England and honorary senior clinical lecturer at the University of Birmingham. His main clinical and research interests include complex spine surgery, including at the craniovertebral junction, Chiari and syringomyelia, neuro-oncology, craniofacial surgery, hydrocephalus, moya-moya, inborn errors of metabolism and congenital conditions, including dysraphism. Guirish is a keen educator and is the West Midlands regional neurosurgery training programme director and chair of the West Midlands surgical training committee for neurosurgery. He is also faculty for the courses run by the European Society of Paediatric Neurosurgery, the International Society of Paediatric Neurosurgery and the paediatric neurosurgery section of the World Federation of Neurosurgical Societies.

STAVROS STIVAROS



Stavros Stivaros is Head of Paediatric Neuroradiology at the Royal Manchester Children's Hospital, one Europe's largest children's hospitals. He is Professor of Paediatric Neuroradiology and Translational Imaging at the University of Manchester, where he leads the paediatric multi-parametric imaging research group. He has a special interest in intracranial haemo/hydrodynamics and the imaging of related disorders. His work on artificial intelligence techniques to fuse and analyse anatomical, physiological and functional imaging in specific disease groups, such as hydrocephalus and children's brain tumours has won international recognition at both the Children's Tumour Foundation as well as the International Society of Paediatric Neuro-oncology.

MARCUS STOODLEY



Professor Stoodley is head of Neurosurgery at Macquarie University Hospital and the Faculty of Medicine and Health Sciences. He graduated with honours from the University of Queensland Medical School. After completing neurosurgery training in Australia, he undertook further subspecialty training in vascular neurosurgery at Stanford University and the University of Chicago in the United States. Professor Stoodley heads the neurosurgery research team at Macquarie University. This is one of the largest neurosurgery research groups in Australasia, with world-leading research efforts in syringomyelia and CSF physiology, and in the development of new treatments for brain arteriovenous malformations. He has produced more than 100 publications and has supervised over 15 research students. In 2012, Professor Stoodley was awarded the John Mitchell Crouch Fellowship by the Royal Australasian College of Surgeons, the premier surgical research award of the RACS.

SOCIAL PROGRAMME

TUESDAY 17TH JULY 2018

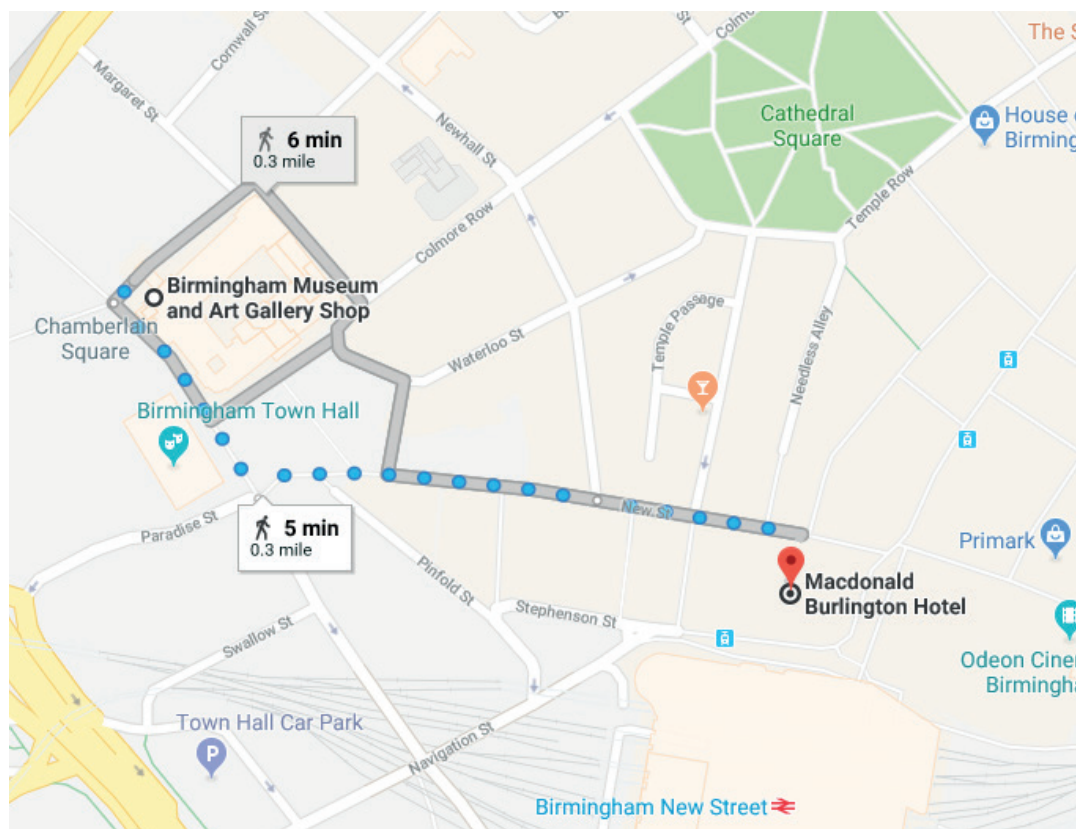
19:00 – Welcome drinks reception and buffet at the Macdonald Burlington Hotel in the Horton Suite

WEDNESDAY 18TH JULY 2018

Free evening

THURSDAY 19TH JULY 2018

Symposium dinner at the Birmingham Museum & Art Gallery
19:00 – arrival drinks
20:00 – seated for dinner



CONFERENCE DINNER MENU THURSDAY 19TH JULY 2018

STARTER

Rose of melon, chilled raspberry soup finished with poached fruits and Greek yogurt

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MAIN

Pan seared breast of chicken, Parma ham and sage mousse served with shoestring vegetables, dauphinoise potatoes and red wine reduction

OR

Mediterranean vegetables and Parmesan Wellington

~

DESSERT

Brioche bread and butter pudding laced with lemon curd and sauce Anglaise

Tea, coffee and handmade truffles

If you have not already, please ensure that you notify a member of the events team ASAP of any special dietary requirements.

ACADEMIC PROGRAMME

DAY 1 – TUESDAY 17TH JULY 2018

08.30 Registration, refreshments and exhibition

SESSION 1: INTRODUCTION AND BACKGROUND TO MEETING

09.45	Welcome	Rebecca Dodwell-Pitt
09.50	Introduction	Graham Flint
10.10	INVITED LECTURE: From Long Island to Birmingham – How far we have come and where we are going	Paolo Bolognese

SESSION 2: STANDARDISATION

10.30	THE BERNARD WILLIAMS MEMORIAL LECTURE: How can we define syringomyelia and measure results of its treatment?	Jörg Klekamp
11.00	INVITED LECTURE: Managing Chiari and syringomyelia – Who, how, when and why?	Andrew Brodbelt
11.30	INVITED LECTURE: Conferences, publications and databases – How can we better learn from each other?	Palma Ciaramitaro
12.00	FREE PAPERS	
F1	Recommended investigations for Chiari type I malformation	Sharon Tam
F2	Surgeons singing from the same hymn sheet – A proposal for the preoperative investigation of patients with Chiari malformation	Nicholas Haden
F3	Proposal to establish a UK Chiari I registry (UK-CIR)	Rory Piper
F4	Syringomyelia and Chiari in the context of European Reference Networks – The challenge of SYRENET	Palma Ciaramitaro

13.00 Lunch, refreshments and exhibition

SESSION 3: CHIARI MALFORMATIONS: SURGICAL AND ON-SURGICAL MANAGEMENT

14.00	INVITED LECTURE: Non-Valsalva headaches, pre- and post-operative – Causes and management	Anita Krishnan
14.30	FREE PAPERS:	
F5	Surgical treatment of syringomyelia associated with Chiari malformation type 1. Analysis of surgical treatment of 124 patients	Andrey Zuev
F6	Minimally Invasive Subpial Tonsilectomy (MIST) and reconstruction of the cisterna magna in the treatment of Chiari malformation type 1 with syringomyelia	Hao Li
F7	Chiari Malformation with or without syringomyelia prospective study – Post-surgery versus conservative long-term outcome in 760 adults	Palma Ciaramitaro
F8	Symptom outcome after craniovertebral decompression for Chiari type 1 malformation without syringomyelia	Joshua Pepper
F9	Re-operation in Chiari patients – The role of the Chiari plate	Francisca Ferreira
F10	Chiari malformation type 1 – A review of literature to compare bony posterior fossa decompression with and without duraplasty	Sharon Tam

16.00 Refreshments and exhibition

16.30	INVITED LECTURE: Beyond the knife and pills	Fiona Adams
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SESSION 3: CONTINUED

17.00	FREE PAPERS:	
	F11	A shunt procedure for post-traumatic and tuberculous syringomyelia treatment Hao Li
	F12	Surgical revision of craniovertebral decompressions - A retrospective review Gopiga Thanabalasundaram
	F13	Arachnoid webs - Clinical and radiological outcomes. Athan Chawira
	F14	Paediatric endoscopic third ventriculostomy - Long-term outcomes in Chiari and syringomyelia Dhruv Parikh
18.00	Close of academic proceedings	
19.00	Welcome reception - Horton Suite, Burlington Hotel	



Don't forget to tweet us during the event:

#SCIS2018@academia_uk

#SCIS2018@2018_symposium

ACADEMIC PROGRAMME

DAY 2 – WEDNESDAY 18TH JULY 2018

08.30 Registration, refreshment and exhibition

SESSION 4: OPERATIVE TECHNIQUES – HOW BEST TO INTERVENE FOR CHIARI

09.00	Get it right first time – The challenge of revisional craniovertebral decompression	Marcus Stoodley
09.20	Chiari malformation type 1– Is it nature's protective air bag?	Atul Goel
09.40	Is craniovertebral decompression the correct procedure to treat Chiari malformation	Guirish Solanki
10.00	Basilar invagination – Decompression, fusion or both?	Jörg Klekamp
10.20	The long-term outcome from occipito-cervical fusion for Ehlers-Danlos hypermobility	Paolo Bolognese
10.40	Moving upstream – The role of venous stenting	Nicholas Higgins
11.00	Refreshments and exhibition	
11.30	Round table discussion	Graham Flint Andrew Brodbelt

SESSION 5: HINDBRAIN-RELATED AND PRIMARY SPINAL SYRINGOMYELIA

12.00	FREE PAPERS:	
F15	Surgical treatment of syringomyelia associated with adhesive arachnoiditis and post-traumatic syringomyelia	Andrey Zuev
F16	Clinical study of section of the filum terminale in the treatment of tethered spinal cord with syringomyelia	Yong Liu
F17	Pitfalls in Chiari malformation type 1 treatment – Possible causes of surgical failures	Marika Furlanetto
F18	Preventing CSF leaks using a pericranial graft during Chiari decompression	Shankar Gopinath
13.00	Lunch, refreshments and exhibition	

SESSION 6: CASE CONSULTATIONS

14.00	Delegates are invited to present anonymised cases, which are clinically challenging or otherwise instructive, to panel of international experts	
16.00	Close of academic proceedings	

ACADEMIC PROGRAMME

DAY 3 – THURSDAY 19TH JULY 2018

08.30 Registration, refreshment and exhibition

SESSION 7: PAEDIATRIC ASPECTS

09.00	INVITED LECTURE: The importance of a tailored approach to the management of Chiari and syringomyelia in children	Christopher Parks
09.30	INVITED LECTURE: Experiences of the full decompression in Chiari 1	Shailendra Magdum
10.00	FREE PAPERS:	
F19	Posterior calvarial distraction for children with complex craniosynostosis, Chiari malformation type 1 and syringomyelia – A 10 year experience in a UK specialist centre	Jonathan Thant
F20	Surgical treatment of paediatric Chiari I malformation – Foramen magnum decompression with dura left open, in 70 patients	Arthur Kurzbuch
F21	Chiari I malformation with significant motor and autonomic dysfunction in a toddler	Mohammad Naushahi
F22	Chiari I malformation – Should we operate on pictures or children? Proposal of a diagnostic and therapeutic flow chart based on the retrospective analysis of 630 mono-institutional cases	Laura Valentini
11.00	Refreshments and exhibition	

SESSION 8: INVESTIGATIONS/CLINICAL ASPECTS

11.30	FREE PAPERS:	
F23	Magnetic Resonance Imaging quantification of morphological parameters in Chiari malformation – A retrospective study.	William Fuell
F24	Hindbrain herniation in hypermobile Ehlers-Danlos syndrome patients	Francis Smith
F25	Neurophysiological and neuroradiological correlates in patients with syringomyelia and Chiari malformation – A central motor conduction time along the phrenic nerve and fibre tracking study	Palma Ciaramitaro
F26	Is cerebrospinal fluid pleocytosis a good indicator of post-operative meningitis following craniovertebral decompression?	Graham Flint
F27	Chiari type 1 malformation related blackouts	Ahmed Elhabel
F28	Presenting symptoms of Chiari malformation as seen from the patient's perspective	Graham Flint
13.00	Lunch, refreshments and exhibition	
14.00	INVITED LECTURE: Chiari and syringomyelia – Where next with imaging	Stavros Stivaros

SESSION 9: ANATOMY/PATHOLOGY/MODELLING

14.30	FREE PAPERS:	
F29	The mechanical role of oedema in cavity formation	Jenny Venton
F30	Computer simulation of syringomyelia in brachycephalic dogs	Serge Cirovic
F31	Syringomyelia pathology – Insights from animal models and ultrastructural studies	Magdalena Lam
F32	Histological analysis of arachnoid features in Chiari I malformation	Jörg Klekamp
F33	Posterior fossa volume of Chiari malformation – Is it funnel narrowing?	Yong Liu
F34	Assessing human brain tissue slices derived from cerebellar tonsillar tissue in Chiari patients, as a laboratory based injury model	Jon Sen
16.00	Refreshments and exhibition	
16:30	THE EDWARD OLDFIELD MEMORIAL LECTURE: Current understanding of the pathophysiology of syringomyelia	John Heiss
17.30	Close of academic proceedings	
19.00	Gala dinner – Birmingham Museum & Art Gallery	

ACADEMIC PROGRAMME

DAY 4 – FRIDAY 20TH JULY 2018

09.00 Registration, refreshments and exhibition

SESSION 10: OBSTETRIC ASPECTS

09.30	INVITED LECTURE: Chiari and syringomyelia; pregnancy and labour; evidence and anecdote	James van Dellen
10.00	FREE PAPERS:	
F35	Management of pregnancy and delivery in women with Chiari malformation type I and/or syringomyelia – A variability survey	Steven Knafo
F36	Pregnancy and labour care for women with Chiari malformation – A case series	Adikarige Silva
10.30	ROUND TABLE DISCUSSION: Obstetric aspects of Chiari and syringomyelia	Marcus Stoodley & James van Dellen
11.00	Refreshments and exhibition	

SESSION 11: VETERINARY ASPECTS

11.30	FREE PAPERS	
F37	MRI biomarkers for canine Chiari malformation-associated pain and syringomyelia	Michaela Spiteri
F38	Vestibular signs, autonomic dysfunction and dysphagia might occur in adult dogs with syringobulbia	Baye Williamson
F39	Persistent fontanelles in Chihuahuas.	Anna-Mariam Kiviranta
12.15	THE ANN CONROY MEMORIAL LECTURE: One health – Learning from our best friends	Clare Rusbridge
12.45	Closing remarks	
13.00	Lunch, refreshments and exhibition	

INVITED LECTURE SUMMARIES

A number of our invited speakers have kindly provided summaries of their presentations

FROM LONG ISLAND TO BIRMINGHAM – HOW FAR WE HAVE COME AND WHERE WE ARE GOING?

Paolo Bolognese

The topics concerning Chiari I Malformation (CMI) have been the subject of discussions and polarizing controversies over the course of many years. During the last decade this vibrant debate has caused a slow shift of opinions among many of the opinion leaders in the field. In order to get a detailed map of the current CMI Weltanschauung among the trendsetters, 100 recognized experts in the field, from 4 Continents, were contacted to answer an ad hoc questionnaire, as a preface to the XXIX Conference of the American Chiari and Syringomyelia Alliance Project (2017, Long Island NY). We received replies from 63 experts; 60 of the responders were neurosurgeons, with a collective surgical experience of more than 15,000 CMI cases. The questionnaire contained 90 questions and was run through the Survey Monkey platform. The first part of the Questionnaire (Questions 1–51) revolved around different CMI topics: pathophysiology, epidemiology, symptomatology, and comorbidities. The second part of the Questionnaire (Questions 52–90, with multiple choice format) focused on surgical management, surgical techniques, and professional profiles. Some surprising facts emerged from the analysis of the results of the first part of the questionnaire (pathophysiology, epidemiology, symptomatology, and comorbidities): 1) a large majority of the replies showed a high level of convergence and consensus on several topics; 2) a number of topics which had been considered controversial as recently as 10 years ago are now regarded as mainstream by many; 3) 88.5% of the responders rejected the validity of the so-called “5mm rule”. The results of this questionnaire not only gave us a current roadmap of our respective opinions and ideas about this disorder but, through the unexpected convergence of consensus about several topics, it is also giving us many building blocks to start the new redefinition and re-classification effort for this disorder.

THE BERNARD WILLIAMS MEMORIAL LECTURE – HOW CAN WE DEFINE SYRINGOMYELIA AND MEASURE RESULTS OF ITS TREATMENT?

Jörg Klekamp

Syringomyelia is considered as a fluid filled cavitation inside the spinal cord. However, there is no agreement on how syringomyelia should be differentiated from other cystic pathologies of the spinal cord. Syringomyelia is the result of a long disease process. Good short-term results of surgical treatment are important for surgeons and patients. However, the real criterion of successful treatment has to be whether the neurological disease can be controlled long-term. But how should long-term results be documented? How long do we need to follow patients before we may consider their condition safe? Among 3206 patients with spinal cord pathologies, identified between 1991 and 2015, 2276 patients demonstrated cystic features. Syringomyelia was differentiated from cystic intramedullary tumours, gliopendymal cysts, myelomalacias and dilatations of the central canal by clinical and radiological criteria. The diagnosis of syringomyelia should be reserved for patients with a fluid filled cavity in the spinal cord related to either a disturbance of CSF flow, spinal cord tethering or an intramedullary tumour. For patients in whom such a relation cannot be established, the diagnosis of syringomyelia should be withheld. Treatment of syringomyelia requires us to treat the underlying cause. Long-term results of surgical procedures and the natural history should be analysed by determining progression-free survival rates to account for varying follow-up times. If different treatment modalities are to be compared, such statistics will reveal differences within 5 years.

CONFERENCES, PUBLICATIONS & DATABASES – HOW CAN WE BETTER LEARN FROM EACH OTHER?

Palma Ciaramitaro

In the rapidly changing world of medicine and its specialties, continuing medical education is considered an essential requirement. Live events, such as conferences, workshops and symposia provide unique learning opportunities that we cannot find anywhere else. Conferences enable healthcare professionals to keep up-to-date with important research, learn directly from experiences, share best practice and develop new skills and techniques. Furthermore, many conference attendees will make contacts that they would not be able to make through other means, such as social networking. Evidence based medicine remains a hot topic for clinicians, public health practitioners and the public, with the use of current best evidence in making decisions about the care of individual patients. This practice involves the integration of individual clinical expertise and the best available external clinical evidence obtained from systematic research (especially from patient centred clinical research) into the accuracy and precision of diagnostic tests, the power of prognostic markers and the efficacy and safety of therapies. Although now established practice, evidence based medicine remains a relatively young discipline, whose positive impacts are just beginning to be validated. It will continue to evolve. The Council of the European Union Recommendation on Rare Diseases (2009) supported the development of patient registers and databases in the area of rare or low prevalence and complex diseases. Patient registry is the best way of pooling data to achieve a sufficient sample size for epidemiological and/or clinical research. Indeed, registries serve as a recruitment tool for the launch of studies focusing on disease aetiology, pathogenesis, diagnosis or therapy. According to the Digital Health programs promoted by European Union, an e-health solution, with validation and implementation of a web platform, is presented -: the CRESSC Database, with clinical research focused on syringomyelia and Chiari malformation.

INVITED LECTURE SUMMARIES

CLINICAL ASPECTS – BEYOND THE KNIFE AND PILLS

Fiona Adams

Many patients with Chiari malformations, as well as some with syringomyelia, may elect to cope with their symptoms and resultant disabilities, rather than undergo a neurosurgical operation. Others, who do have surgery and who, hopefully, gain some benefit, may still be left with some of their original symptoms. Occasionally surgery causes complications, with new symptoms developing. Referral to a neurology service, or a pain clinic, is a common pathway for such individuals, following which various drug regimes are commonly tried. There are, however, a number of other therapies that can be applied, with benefit. Physiotherapy may assist in the control of subjective balance disturbances or with alleviation of vestibular symptoms, as well as compensating for motor deficits and assisting functional restoration and enabling the client to regain some exercise ability. Speech and language therapy may help the patient who presents with bulbar symptoms, such as dysphonia or dysphasia. Diet and exercise advice should help with weight loss when this is aggravating matters. Occupational therapy can assist in many areas including applying desensitisation techniques may help with management of chronic pains and sensory disturbances, affecting various parts of the body. Management of chronic fatigue, a common complaint with Chiari and syringomyelia patients, can yield great benefits. So, too, can the alleviation of anxieties and the inevitable psychological distresses that accompany living with these chronic neurological disorders. Chiari malformations and syringomyelia can have life-altering effects on people and, beyond any surgical or pharmacological treatments offered by neurosurgeons, neurologists and pain relief specialists, the Therapies can help the individual rebuild norms in his or her life.

CHIARI MALFORMATION TYPE 1 – IS IT NATURE'S PROTECTIVE AIR BAG?

Atul Goel

Understanding that atlantoaxial instability is the cause of Chiari malformation (CM), the author treated 275 patients using atlantoaxial stabilization during the period from January 2010 to December 2017. Surgery was aimed at segmental arthrodesis. Twenty patients had been treated earlier using foramen magnum decompression and duraplasty. According to the extent of their functional capabilities, patients were divided into 5 clinical grades. On the basis of the type of facet alignment and atlantoaxial instability, the patients were divided into 3 groups. Type I dislocation was anterior atlantoaxial instability wherein the facet of the atlas was dislocated anterior to the facet of the axis. Type II dislocation was posterior atlantoaxial instability wherein the facet of the atlas was dislocated posterior to the facet of the axis. Type III dislocation was the absence of demonstrable facet malalignment and was labelled as "central" atlantoaxial dislocation. All patients were treated with atlantoaxial plate and screw fixation using techniques described in 1994 and 2004. Foramen magnum decompression or syrinx manipulation was not performed in any patient. Occipital bone and sub axial spinal elements were not included in the fixation construct. On the basis of outcomes in this study, it appears that the pathogenesis of CM, with or without associated basilar invagination and/or syringomyelia, is primarily related to atlantoaxial instability. The data suggest that the surgical treatment in these cases should be directed toward atlantoaxial stabilization and segmental arthrodesis. Except in cases in which there is assimilation of the atlas, inclusion of the occipital bone is neither indicated nor provides optimum stability. Foramen magnum decompression is not necessary and maybe counter-effective in the long run.

BASILAR INVAGINATION – DECOMPRESSION, FUSION OR BOTH?

Jörg Klekamp

Basilar invagination is a rare craniocervical malformation, which may lead to neurological deficits related to compression of the brainstem and upper cervical cord, as well as instability of the craniocervical junction. 124 patients with basilar invagination were encountered. The clinical courses were documented with a score system for individual neurological symptoms for short-term results after 3 and 12 months. Long-term outcomes were analysed with Kaplan-Meier statistics. Patients with (n = 55) or without (n = 69) ventral compression were distinguished. 56 patients declined an operation, while 68 patients underwent surgery. Surgical management depended on the presence of ventral compression and segmentation anomalies between occiput and C3, signs of instability and presence of caudal cranial nerve dysfunctions. Of 28 patients without ventral compression, 19 underwent a foramen magnum decompression for the associated Chiari I malformation only, while 9 underwent decompression and fusion. Among 40 patients with ventral compression, 29 patients required a decompression and fusion, while 11 were treated by decompression only. Within the first postoperative year neurological scores improved for all symptoms in each patient group. In the long-term, postoperative deteriorations were related exclusively to instabilities either becoming manifest after a foramen magnum decompression in 3, or as a result of hardware failures in 2 patients. The great majority of patients with basilar invagination report postoperative improvements with this management algorithm. Most patients without ventral compression can be managed by foramen magnum decompression alone. The majority of patients with ventral compression can be treated by posterior decompression, realignment and stabilization alone, reserving anterior decompressions for patients with profound, symptomatic brainstem compression. Patients undergoing decompression only require close post-operative follow-up to rule out post-operative C1/2 instabilities

CONDYLAR SCREW FIXATION IN OCCIPITO-CERVICAL FUSIONS

Paolo Bolognese

Surgical fixation at the craniovertebral junction is indicated in a number of clinical pathologies. Condylar screw fixation has been recently suggested and validated as a rescue technique and an alternative to the conventional configuration, whenever former surgical bone removal along the supraocciput makes the anchoring of the plate technically difficult. Cadaveric investigations and a limited number of case studies using occipital condyle (C0) fixation have been published so far, in both the adult and paediatric population. The challenging dissection of C0-1, concerns about possible complications, and the overall modest number of occipitocervical fusions required have thus far prevented the acquisition of large surgical series utilizing occipital condylar screws. In this IRB approved study (IRB #13-655B), we present our single-surgeon experience accumulated in 250 cases of occipitocervical fusions using occipital condylar screws, over a period of 8 years. Only two direct complications from condylar screw insertion occurred in this study, and resulted in a complete reversal of the deficit. At short and long follow-up, the subjects of the condylar cohort have demonstrated good levels of postsurgical clinical improvement, solid bone fusions, and a low amount of local discomfort from the hardware.

INVITED LECTURE SUMMARIES

CHIARI & SYRINGOMYELIA – WHERE NEXT WITH IMAGING

Stavros Stivaros

Until the relatively recent past, the main focus of development in brain imaging has been on either greater tissue contrast or improvements in resolution. We now have imaging studies routinely producing anatomical imaging with a resolution of less than 1mm³. Sometimes, however, simple anatomy is not enough and we find that the snapshot of an anatomical image has a relatively poor diagnostic efficacy. Fortunately, with the advent of multi-parametric brain imaging, we are now able to undertake imaging studies that also inform on function and physiology. The imaging assessment of hydrocephalus and Chiari have benefited from just such multi-parametric imaging assessments. This lecture will look at the development of neuroradiology and examine the techniques that have helped to unravel some of the physiological aspects of the haemo- and hydro-dynamics associated with CSF production and flow, including phase contrast MRI, diffusion and perfusion imaging. In addition we will explore how these same imaging techniques can be used to assess pathophysiology in patients, aid in diagnosis and inform on outcome, on an individual patient basis.

THE EDWARD OLDFIELD MEMORIAL LECTURE – CURRENT UNDERSTANDING OF THE PATHOPHYSIOLOGY OF SYRINGOMYELIA

John Heiss

Solving the riddle of syringomyelia pathogenesis requires an explanation of how fluid, either CSF or biochemically identical to CSF, is entrained in the spinal cord central canal, accumulates to form a syrinx, and distends the spinal cord. During syrinx expansion the volume of fluid entering the central canal (CSFin) must exceed that leaving that structure (CSFout). The late Dr Edward Oldfield and I published a study comparing transit of myelogram dye into and out of syringes among patients with syringomyelia associated with either Chiari I malformation, spinal subarachnoid lesion, or haemangioblastoma, an intramedullary tumour. We found that more dye entered syringes associated with Chiari I malformation or spinal lesions than those associated with intramedullary tumours, suggesting that syringomyelia associated with spinal subarachnoid space obstruction was maintained by fluid passing from the spinal subarachnoid space, through the spinal cord, and into the syrinx, whereas intramedullary tumours maintained their syringes through intra-tumour fluid production. Dr Oldfield and colleagues earlier published a study using contrast-enhanced FLAIR imaging to show that haemangioblastomas of the cerebellar hemisphere and spinal cord produce fluid that passes enters the surrounding extravascular spaces, creating oedema and eventual cyst or syrinx formation. Because internal fluid production has not been shown in syringes associated with spinal subarachnoid obstruction, we propose another mechanism in which enlarged subarachnoid pressure waves drive an external source of syrinx fluid, CSF, into the spinal cord to form a syrinx.

PREGNANCY AND LABOUR – EVIDENCE AND ANECDOTE

James van Dellen

In the intriguing topic of Chiari and syringomyelia there is, as yet, an unexplored aspect and that is the circumstances surrounding pregnancy and parturition. There is a female dominance with Chiari and there would be understandable concern, in female patients diagnosed with Chiari or syringomyelia, regarding their falling pregnant and the effects that this would have upon them. The default advice is largely to consider family planning very carefully, and some women are even advised to avoid falling pregnant altogether. There are many who would not be aware that they have Chiari or syringomyelia and who have had children before they were first diagnosed. There are also those who are, very exceptionally, diagnosed during pregnancy and, extremely rarely, during parturition. Such instances are without recorded scientific case reports of any adverse events but more likely simply anecdotal statements of concern. There are case reports and small series of reports on delivery practices with Chiari/syringomyelia mothers-to-be, with the vast majority taking a default position of advising elective Caesarean section. Again there are anecdotal case descriptions, not scientifically recorded, of 'misadventures' with parturition and pain control. Litigation consequences are the main concern. The conundrum persists but, as yet, very few substantive reports exist; a conundrum because a normal vaginal delivery might reasonably be regarded as the ultimate Valsalva manoeuvre. In terms of current views regarding the mechanisms of origin and symptom production from Chiari and syringomyelia, we must ask ourselves, how do pregnancy and parturition stack up against these theories?

INVITED LECTURE SUMMARIES

THE ANN CONROY MEMORIAL LECTURE: ONE HEALTH; LEARNING FROM OUR BEST FRIENDS

Clare Rusbridge

Syringomyelia is an increasingly common diagnosis in veterinary medicine and most frequently associated with Chiari-like malformation (CM). Eponymous terms are discouraged in veterinary medicine, although not without precedent (for example Horner's syndrome and Wallerian degeneration). However this label, chosen in a round table discussion was considered less restrictive than an anatomical description such as hindbrain herniation because the latter could prove to be too simplistic or inaccurate in the future. This was a wise decision because, in the 20 years since the first description in the dog, our understanding of CM has morphed from a description of cerebellar herniation (dogs do not have cerebellar tonsils) to a complex developmental malformation of the skull and craniocervical vertebrae characterized by a conformational change and overcrowding of the brain and cervical spinal cord, particularly at the craniospinal junction. CM is associated with miniaturisation and brachycephalism and is prevalent in several toy dog breeds particularly Cavalier King Charles Spaniel and Chihuahua to a lesser extent, the fashionably popular French Bulldog and Pug dog. Perhaps surprising, many toy breeds with extreme facial foreshortening such as the Pekinese, Japanese chin and Shi Tzu, are not predisposed perhaps reflecting a different skull shape, brain size and genetic heritage. Since the bony and parenchymal changes between and within individuals in each breed are different, CM might be considered an umbrella term with a common tendency towards pain associated with CM and the development of syringomyelia. As in humans, obstruction of the foramen magnum and cerebrospinal fluid channels is pivotal in the pathogenesis of syringomyelia and the variation in phenotype makes this condition similar to brachycephalic obstructive airway syndrome (BOAS) – the most common cause of respiratory distress in companion dogs. Many dogs are affected by both conditions. Although there is a phenotypic variation, this is not as marked as in humans and consequently our canine companions represent a huge resource for understanding the pathogenesis of syringomyelia, diagnosis, natural history of the disease, treatment options, and genetics. Machine learning techniques, (as illustrated in one of the free papers at the symposium), could revolutionise diagnosis providing an objective measure of CM. The principle can be readily applied to humans. Another of the veterinary papers describes computer modelling of syringomyelia from canine MRI. This technique provides insights into the pathogenesis and, in theory, might be developed to model surgical outcome. The hypothesis being that different surgeries can be modelled on a patient in vivo in order to select the most appropriate procedure. As a naturally occurring and long-lived model of syringomyelia and central neuropathic pain that are subject to similar co-morbidities such as obesity and cardiovascular disease, clinical trials in dogs could provide insights into pain management in humans. Studies into the genetics of the condition have shown CM/SM to be a complex trait, which can be late onset with a moderately high heritability that likely involves genes involved in embryonically-active pro-osteogenic signalling pathways. To date, three candidate genes have been identified: SALL-1 (in humans Mutations in the SALL1 gene cause Townes-Brocks Syndrome which can be associated with CM); PCDH17, a CNS cell adhesion molecule; and ZWINT of which expression is associated with neuropathic pain. Thus Chiari malformation and syringomyelia are an excellent examples of One Health since "solutions require everyone working together on all the different levels" (quotation from William Karesh credited with coining the modern usage of the term via journalist Rick Weiss Washington Post). And this certainly applies in this case to 'Man and his Dog'.

FREE PAPER ABSTRACTS

F1 RECOMMENDED INVESTIGATIONS FOR CHIARI TYPE I MALFORMATION

Foroughi M, Tam S.

INTRODUCTION

The prevalence of Chiari type I malformation (CM-I) has been estimated to be between 1% in adults and up to 3% of the paediatric population. The majority are asymptomatic and not associated with any underlying pathology, such as hydrocephalus, syringomyelia or C1/C2 instability. The aim of this review was to appraise critically the benefits of various imaging modalities for diagnosing this malformation and its associated pathologies.

METHODS

The authors reviewed the pertinent literature on the neuroradiology of the CM-I, giving special regard to magnetic resonance imaging (MRI) of the entire spine and brain, constructive interference in steady state (CISS) image MRI sagittal T2, Cine-MRI, and C1 or C2 flexion and extension imaging. A literature search was performed using PubMed and Embase databases and the search was focused on imaging techniques used to diagnose CM-I.

RESULTS

MRI of the entire spine and brain enables one to assess for hydrocephalus, syringomyelia and tethered cord. Cine-MRI provides information on CSF pulsatility at the craniocervical junction, while CISS image MRI sagittal T2 allows better-quality visualisation of CSF around the cerebellum and tonsils. Both techniques provide useful additional details but are not mandatory CM-I diagnostic tools. CM-I associated C1/C2 instability is not commonly seen in the UK but if suspected the use of C1/C2 flexion and extension X-rays and CT scan is recommended.

CONCLUSION

Asymptomatic CM-I hindbrain hernias without impaction do not need further investigations. Symptomatic or significant impaction CM-I hindbrain hernias should be investigated using MRI brain and entire spine, preferably using thin cut high resolution T2 weighted images of the craniocervical junction. Flexion and extension x-rays or CT scans are indicated only when instability of C1/C2 region is suspected. Cine MRI is not essential but can provide benefits for assessment of CSF flow.

F2 SURGEONS SINGING FROM THE SAME HYMN SHEET - A PROPOSAL FOR THE PRE-OPERATIVE INVESTIGATION OF PATIENTS WITH CHIARI MALFORMATION

Mediratta S, Haden N.

INTRODUCTION

The reliability provided by doing the same thing in the same way, for comparable patients, is increasingly recognised and respected in all surgical fields. NHS England has invested significantly in its GIRFT (Getting it Right First Time) project, to identify outliers in surgical practice and to help focus efforts on delivering consistency and efficiency. A challenge remains, with any low volume surgery, with regard to just what is "right"? Discussions at meetings of the British Syringomyelia-Chiari Group (BSCG) reveal how even a group of surgeons dedicated to the care of patients with these pathologies can be inconsistent in their approach to investigation, assessment and anticipated benefits of their proposed management. Local neurosurgical practice in the south-west of England has changed over time. For example, CSF flow studies, not previously an essential component of the local work-up of patients, are now seen as having specific and independent value.

METHODS

This study aimed to review current UK practices and to assess literature evidence, in order to propose a set of guidelines for discussion and subsequent adoption into neurosurgical practice. An online questionnaire was sent to all members of the BSCG, to assess current practice with respect to the investigation of patients presenting with a Chiari malformation - with a request to forward this questionnaire to any colleagues locally, who also undertake such procedures. The data was collated so that current practice could be summarised. A literature review of published practice and evidence was also undertaken and comparison between this and current practice was made.

RESULTS

A proposed set of guidelines for consistent, comparable and complete assessment of patients presenting with Chiari malformations has been prepared for discussion.

F3 PROPOSAL TO ESTABLISH A UK CHIARI I REGISTRY (UK-CIR).

Piper RJ, Magdum SA, Jayamohan J.

BACKGROUND

The evidence base for managing Chiari I malformation (CM-I) currently consists of relatively small, single-centre and/or retrospective studies. The lack of high quality data poses challenges to our understanding of the condition and its optimal management, as well as the creation of reliable clinical guidelines and planning for service provision. We therefore propose to establish a UK Chiari I Registry (UK-CIR).

OBJECTIVES

The objective of the UK-CIR is to prospectively acquire UK-wide data for patients with CM-I in order to: 1) report the current UK-wide epidemiology of CM-I; 2) determine the number of patients treated for CM-I in the UK and to examine variances in practice; 3) investigate the short and long-term outcomes following conservative and surgical managements of CM-I.

PROPOSED METHODS

The proposed UK-CIR is a UK-wide, multicentre, prospective and observational data registry. All UK neurosurgical units will be invited to participate. The UK-CIR will follow the British Neurosurgical Trainee Research Collaborative (BNTRC) model for multicentre collaborative research. All neurosurgically-naïve patients (paediatric and adults) with CM-I are eligible to be enrolled. Anonymous data will be directly recorded into an electronic, online database system. Local investigators will have full access to their local data in order to facilitate local audit, but not the data of other participating units. The data collected in the UK-CIR is divided into four phases: 1) demographic data; 2) presentation data; 3) surgical data; 4) outcome data. Data from phases 1 and 2 will be collected preoperatively. Data for phases 3 and 4 will be collected postoperatively at 1, 5 and 10yrs. The UK-CIR will initially be piloted in at least two UK neurosurgical units before it is offered out nationally.

FREE PAPER ABSTRACTS

F4 SYRINGOMYELIA AND CHIARI IN THE CONTEXT OF EUROPEAN REFERENCE NETWORKS: THE CHALLENGE OF SYRENET.

Ciaramitaro P, Peretta P, Valentini L, Morar S, Parker F, Poca MA, Sauquillo J, Celada C, Gilanton M, Flint G.

INTRODUCTION

Based on European Union directive 2011/24/EU, on the application of patients' rights in cross-border healthcare, European Reference Networks (ERNs) require 10 healthcare providers, across 8 member states to: 1) facilitate European co-operation on highly specialized healthcare systems; 2) facilitate concentration of expertise for providing high quality healthcare; 3) reinforce research, epidemiological surveillance and training. The main challenge of SYRENET (Syringomyelia and Chiari European Network) will be to decrease existing inequalities in access to high quality health care for syringomyelia and Chiari.

METHODS

28 centres, from 10 European Countries (Belgium, Bulgaria, Czech Republic, France, Italy, Norway, Portugal, Romania, Spain, United Kingdom) and 1 international partner country (US) are the pilot centres. Key aims are to: 1) create an on-line database, to improve the comprehensive data collection; 2) identify expertise centres, for information collection, medical training, research, and dissemination of information; 3) involve patients, through national syringomyelia and Chiari associations.

RESULTS

CRESSC (www.cressc.org) is an international, multilingual, public and free web platform, easily accessed by clinicians and patients, on multiple devices (computers, smartphones, tablets). Key performance indicators, quantitative and qualitative, such as accessibility of health services, appropriateness, efficacy, safety and quality were identified by the pilot centres of the Network.

DISCUSSION

A shared web platform could reduce the existing variability in diagnostic practices, clinical care and prevention strategies, internationally. A multidisciplinary care model, involving experts in neurology, neurosurgery, neurosciences, genetics, veterinary medicine and drug treatment, will help to identify diagnostic targets and should stimulate innovative therapeutic approaches.

CONCLUSIONS

SYRENET is an international, multidisciplinary, open, collaborative network, based on a high level of synergy, integration and complementarity, involving expertise centres, patient associations and researchers. An action plan for future collaborative and interdisciplinary research is proposed, backed by Horizon 2020 funding programs and involving European and other international partners.

F5 SURGICAL TREATMENT OF SYRINGOMYELIA ASSOCIATED WITH CHIARI MALFORMATION TYPE 1. ANALYSIS OF SURGICAL TREATMENT OF 124 PATIENTS.

Zuev A, Pedyash N, Epifanov D, Lebedev V, Ghodiwala T.

INTRODUCTION

The purpose of the study was to analyse the results of treatment of patients with syringomyelia associated with Chiari malformation type 1.

METHODS

During the period from 2013 to 2017 we treated 221 patients with syringomyelia associated with Chiari malformation type 1. Of these, 124 patients were treated surgically, all operations being performed by one surgeon. All patients underwent clinical investigation before surgery and thorough observation was carried out post-operation. All patients underwent MRI investigation in the early postoperative period, to assess the adequacy of decompression. Further, clinical and MRI examinations were carried out at 4 months and 1 year after surgery, for most patients, and annually thereafter.

RESULTS

At 4 months after surgery the syringomyelia cavity disappeared in 19 patients (15%), decreased in 89 (72%) and remained unchanged in 16 patients (13%). At 1 year after the operation, the clinical condition had either improved or remained unchanged in 120 patients (97%). In 4 patients (3%) the syringomyelia continued to progress clinically. Cine-MRI in these cases showed no disturbance of CSF flow at the craniocervical junction and these patients therefore underwent syringo-subarachnoid shunting, resulting in a good clinical outcome. During follow-up we had no relapse of CSF flow obstruction at the craniocervical junction. Early postoperative complications occurred in 4 patients (3%): 1 CSF leak, 1 acute epidural hematoma, and 2 cases of aseptic meningitis. In 11 patients the headache increased initially after surgery but improved after about one month.

CONCLUSION

Sub-occipital craniotomy, followed by duraplasty and restoration of CSF flow at the craniocervical junction, is an effective method of treatment for patients with syringomyelia associated with Chiari malformation type 1

FREE PAPER ABSTRACTS

F6 MINIMALLY INVASIVE SUB-PIAL TONSILLECTOMY (MIST) AND RECONSTRUCTION OF THE CISTERNA MAGNA IN THE TREATMENT OF CHIARI MALFORMATION TYPE 1 WITH SYRINGOMYELIA

Lou Y, Li H, Jin Y, Liu L.

OBJECTIVE

Minimally Invasive Sub-pial Tonsillectomy (MIST) and reconstruction of the cisterna magna, resolves the tonsillar herniation and reconstitutes the cisterna magna, which in turn restores cerebrospinal fluid circulation. Intraoperative ultrasound is used. In this study we assess the outcomes of the procedure in the management of Chiari malformation type 1 with syringomyelia.

METHODS

Between January 2014 and June 2015, 130 patients meet the inclusion criteria. The Chicago Chiari Outcome Scale (CCOS) was used to assess clinical outcomes. Post-operative MRI at 6 months assessed the change in the syrinx. We compared our cohort with a historical control group of 167 cases and a literature group of 378 cases, using the Fisher exact test (significance level: $p < 0.05$)

RESULTS

In our group of 130 cases, 127 patients (98%) had good post-operative CCOS (≥ 11), whereas in 3 cases the score was poor (4-10). The relevant rate of improvement in the historical control group of 167 patients was 82, which is significantly lower ($p = 0.032$). The MRI at 6 months post-operatively showed reduction or complete resolution in all 130 cases.

CONCLUSION

The Minimally Invasive Sub-pial Tonsillectomy and reconstruction of the cisterna magna in patients with Chiari malformation type 1 and syringomyelia has very good clinical and radiological outcomes compared with traditional surgery. It is associated with a low complication rate and emphasizes the importance of reconstructing normal posterior fossa anatomy.

F7 CHIARI MALFORMATION WITH OR WITHOUT SYRINGOMYELIA PROSPECTIVE STUDY: POST-SURGERY VERSUS CONSERVATIVE LONG-TERM OUTCOME IN 760 ADULTS

Ciaramitaro P, Ferraris M, Rosso I, Petrozzino S, Petrillo M, Valentini CM, Costa P, Ducati A, Massaro F.

INTRODUCTION

A longitudinal prospective study was conducted in adults affected by Chiari I syringomyelia complex and Chiari syndrome. The purpose of the study was to evaluate the long term outcome (12-48 months) after surgery (craniocervical decompression with a synthetic patch), compared with conservative management.

METHODS

Between 2010 and 2016, 760 patients (230 male, 530 female, average age 48 ± 16 years), affected by Chiari I malformation, with or without syringomyelia, were admitted to the Inter-regional Expertise Centre for Syringomyelia and Chiari Syndrome (CRESSC) in Turin. Chiari syndrome and Chiari I syringomyelia complex diagnoses were confirmed by clinical and neuroradiological criteria, according to standardized recommendations. Patients were prospectively evaluated, both clinically (MRC, VAS, DN4, Rankin scales) and with neuroimaging (brain and whole spinal cord, cine-mode MRI). In the clinical and neuroradiological follow-up Good Outcome Scores were calculated.

RESULTS

56 patients underwent surgery (15 male, 41 female), average age 47 years (range 19-72). Of these, 26 with Chiari I syringomyelia complex and 15 with Chiari syndrome enjoyed a good clinical outcome (77 and 93% respectively). A good neuroradiological outcome was achieved in all patients in both groups. Of patients treated conservatively, 12(83%) with Chiari I syringomyelia complex had a good neuroradiological outcome but cavity enlargement was seen in the remaining 17%. In the 3 conservatively managed Chiari syndrome patients neither improvement nor worsening was observed.

CONCLUSIONS

In this prospective, long-term study a good outcome was observed after surgical treatment, in terms of functional recovery, in 77-97% of patients with Chiari I malformation, with or without syringomyelia, following careful patient selection. The clinical and neuroradiological outcome with conservative management was also good (83%). These results need to be confirmed with longer follow up (> 5 years) and in wider cohorts of Chiari malformation patients.

F8 SYMPTOM OUTCOME AFTER CRANIOVERTEBRAL DECOMPRESSION FOR CHIARI TYPE 1 MALFORMATION WITHOUT SYRINGOMYELIA

Pepper J, Elhabel A, Tsermoulas G, Flint G.

ABSTRACT. Specific symptom outcome after craniocervical decompression is not well defined in patients with Chiari I malformation. In this study we performed a retrospective review of all patients diagnosed with Chiari I malformation without syringomyelia, who underwent craniocervical decompression in our unit, and assessed the symptoms that improved after surgery. In total, 133 patients were identified with a minimum 2 year follow up. The most common pre-operative symptoms were pressure dissociation headaches (78%), visual disturbances (34%), dizziness/balance disturbances (24%) and blackouts (17%). The symptoms most likely to respond to surgery included headache (72% response, $p < 0.0001$), blackouts (response 87%, $P = 0.0018$) and visual symptoms (58% response, $p = 0.049$).

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F9 RE-OPERATION IN CHIARI PATIENTS -THE ROLE OF THE CHIARI PLATE

Ferreira F, Brodbelt A, Piggot T, Buxton N.

AIM

To investigate patient and surgical factors related to a successful outcome in patients undergoing revision Chiari surgery.

METHODS

Retrospective case note audit, utilizing patient and clinician reported outcome scores, of patients who required revision Chiari surgery in a single institution from 2009. The role of C1 laminectomy, craniectomy size, duraplasty, arachnoid opening, cerebellar tonsillar cauterization, shunting, and the use of the Chiari plate were examined. Outcomes were standardized and compared through Core Outcomes Measures Index (COMI) scores. Where COMI scores were absent, clinic letter outcome descriptions were used.

RESULTS

Seventeen patients were treated for recurrent symptoms. Nine patients had cerebellar slump and 8 of these underwent Chiari plate placement. COMI scores improved in 3 of these, remained stable in 1 and deteriorated in one. COMI score also deteriorated in one patient who was not plated. For three patients undergoing plate placement, only clinic outcome descriptions were available; 1 patient improved and 2 stabilized. Eight patients had undergone a previous, inadequate bony decompression. They underwent a larger craniectomy, with dural opening and tonsillar cauterization. Three of these were worse postoperatively and 4 needed shunting.

CONCLUSION

In revision Chiari surgery, the principles of maintenance of fluid dynamics, and conservative bony decompression are paramount. A Chiari plate can successfully help with treating cerebellar slump. Poor results appear to be mainly associated with the absence of duraplasty and shunt insertion.

F10 CHIARI MALFORMATION TYPE 1 - A REVIEW OF LITERATURE TO COMPARE BONY POSTERIOR FOSSA DECOMPRESSION WITH AND WITHOUT DURAPLASTY

Foroughi M, Tam S.

BACKGROUND

Two main surgical approaches for symptomatic Chiari type I malformation (CM-I) patients are posterior fossa decompression involving craniectomy alone (PFD), and posterior fossa decompression with duraplasty (PFDD). The aim of this review was to outline the indications, advantages and disadvantages of each surgical approach, with guidance regarding surgical decisions.

METHODS

We reviewed pertinent articles, retrieved by searching in the PubMed and Embase databases. Inclusion and exclusion criteria were predefined. Data on the surgical outcomes, complications, re-operations, duration of procedure and the length of hospital stay were compared.

RESULTS

Eighteen articles, containing data on 2,840 paediatric and adult participants, met the inclusion criteria. PFDD was associated with more favourable surgical outcomes. Regarding radiological outcomes, syrinx reduction was observed in 92.3% PFDD vs 12.5% PFD ($p=0.001$). On the other hand, PFD was associated with lower complications rates. The rate of postoperative aseptic meningitis was 6.1% in PFD vs 27.1% ($p=0.027$) in PFDD, and the rate of procedural-related complication was 0.8% in PFD vs 2.3% in PFDD ($p=0.008$). However, PFD was linked to higher rates of re-operation with an odds ratio of 0.15 in PFDD vs PFD ($p=0.002$).

CONCLUSION

Both PFD and PFDD are effective and safe surgical strategies for symptomatic CM-I, associated with posterior fossa volume mismatch and in the absence of hydrocephalus and craniocervical region instability. Bony PFD has a lower complication rate, and seems to be good option when carried out in the paediatric age group, in individuals without major tonsillar impaction, and in the absence of a syrinx. However, these patients should be adequately counselled regarding the requirement for possible further, intra-dural decompression.

F11 A SHUNT PROCEDURE FOR POST-TRAUMATIC AND TUBERCULOUS SYRINGOMYELIA TREATMENT

Lou Y, Jin Y, Liu L.

OBJECTIVE

The surgical treatment of post-traumatic and post-tuberculous syringomyelia is challenging. The authors present their surgical technique of placing a subarachnoid shunt above and below the adhesions with two separate laminectomies in order to restore the subarachnoid CSF circulation.

METHOD

Between 2016 and 2017, we treated 32 cases (20 men and 12 women) with post-traumatic and post-tuberculous syringomyelia. The surgical procedure consisted of two separate midline incisions and laminectomies, rostral and caudal to the adhesions. The normal subarachnoid space was exposed and a tube was inserted in each space. The two tubes were connected in the para-spinal plane. The outcomes were assessed clinically and with MRI

RESULTS

At 12 months after the surgery 26 patients had improvement in their neurological symptoms and the MRI showed a decrease of the syrinxes. Six cases had no change postoperatively, and the MRI showed no significant changes of the syrinxes. Long term follow up is in progress.

Conclusion. Trauma and TB infection of the spinal cord can cause interference of the normal CSF flow, which may lead to syringomyelia. Discrete segment subarachnoid shunting is safe and effective for patients with post-traumatic and post-tuberculous syringomyelia.

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F12 SURGICAL REVISION OF CRANIOVERTEBRAL DECOMPRESSIONS – A RETROSPECTIVE REVIEW

Thanabalasundaram G, Silva A, Tsermoulas G, Flint G.

OBJECTIVE

Craniovertebral decompression is performed for symptomatic Chiari type 1 malformation, with or without syringomyelia. The procedure usually improves Valsalva-related headaches and the syrinx often collapses but in a few patients the symptoms and/or the syrinx persist or recur, in which case revision surgery may be offered. The aim of this study was to examine the indications, technique and outcomes for revision craniovertebral decompression.

METHODS

We retrospectively reviewed all revision craniovertebral decompressions performed in our centre over an eighteen-year period. We collected data on demographics, patient's symptoms and imaging. We analysed the data with descriptive statistics. A total of 35 patients were identified (23 females, 12 males). Twenty patients had isolated Chiari 1 and 15 also had syringomyelia. The follow up period was 1–12 years (mean 3 years).

RESULTS

The mean time to revision was 80 months, range 9–264. For the majority (n=31) this was their 1st revision; for 3 it was their 2nd revision and in one case it was a 3rd revision. Overall, 32 of the 35 patients reported improvement after surgery, including all patients with syringomyelia. The symptom that improved most commonly was headache. Ten of the 15 syringomyelia patients demonstrated radiological resolution of the syrinx post-operatively and another 2 showed reduction in syrinx size. Out of the total group of 32 patients enjoying symptomatic improvement, 10 later developed symptom recurrence, with a mean time to recurrence of 12 months (range 5–24). Nine of these recurrences were in patients with only Chiari; the tenth recurrence was the only instance of such amongst the syringomyelia patients.

CONCLUSIONS

Revision craniovertebral decompression results in early symptomatic relief in most patients with Chiari 1 malformation and, in the medium-term, this benefit is sustained in about two thirds of patients.

F13 ARACHNOID WEBS – CLINICAL AND RADIOLOGICAL OUTCOMES

Chawira A, Buxton N, Pigott T, Brodbelt AR.

BACKGROUND

Arachnoid webs can be challenging to diagnose. The aim of this retrospective, single-centre study was to examine clinical and radiological outcomes in symptomatic patients undergoing arachnoid band division surgery between 2007 and 2017.

METHODS

1046 patient operative records were examined. 15 patients (4 female, 11 male) met the inclusion criteria. An examination of notes, images, and operative reports was conducted. Clinical outcome was stratified into 'worse', 'same', or 'better'.

RESULTS

Median age was 62 years (range 26–84). Median radiological follow-up was 6 years and median clinical follow-up was 2 years. Median duration of pre-operative symptoms was 31 months (range 4–288). Arachnoid bands were identified on myelography or on MRI and confirmed intra-operatively. In the follow-up period, 6 patients (4 in 10) demonstrated a reduction in the volume of their syrinx based on sagittal T2-weighted MR images, with 8 patients (just over half) experiencing either improvement in their symptoms or stability post-operatively. Radiological improvement or stability was observed in 12 patients and was associated with clinical improvement in 3 cases, stability in 4 and worsening in 5 cases.

CONCLUSIONS

Arachnoid webs can be difficult to diagnose and a high index of suspicion is required. Patients with imaging evidence of syrinx improvement may continue to deteriorate. A better understanding of the pathophysiology may help to improve outcome.

F14 PAEDIATRIC ENDOSCOPIC THIRD VENTRICULOSTOMY – LONG-TERM OUTCOMES IN CHIARI & SYRINGOMYELIA

Parikh D, Slator N, Mundil N, Rodrigues D, Walsh AR, Solanki GA.

INTRODUCTION

Endoscopic Third Ventriculostomy (ETV) is an established therapy for obstructive hydrocephalus. We review long-term outcomes at our institution and highlight the use of ETV for the lesser known indication of Chiari malformation and syringomyelia

METHODS

A retrospective review of 139 ETVs performed between 2006 and 2017. The median follow-up was 69 months (range 3–132). Medical records were retrieved from our electronic database and imaging reviewed in our PACS system. Statistical methods performed included a time-to-event analysis (where event = further hydrocephalus procedure) and chi-squared test between observed and predicted success as per ETV Success Score (ETVSS)

RESULTS

130 Patients underwent 139 ETVs. The majority were older than 1 year. The commonest indication was aqueduct stenosis by tumour. In primary procedures the overall ETV success was 74%. The timing of ETV failure showed that 44% occurred in under 30 days and 25% between 30 days and 6 months. ETV success at 6 months was 82%. The predicted ETVSS was 71%, $p=0.59$ on a chi-square test. The percentage survival was evaluated with a Kaplan-Meier curve. ETV success in Chiari & syringomyelia was 9 out of every 10 cases.

CONCLUSIONS

The main discriminators of ETV success are neonatal age and aetiology. ETV alone has been successful in resolving Chiari I and syringomyelia associated with ventriculomegaly.

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F15 SURGICAL TREATMENT OF SYRINGOMYELIA ASSOCIATED WITH ADHESIVE ARACHNODITIS AND POST-TRAUMATIC SYRINGOMYELIA

Zuev A, Pedyash N, Epifanov D, Lebedev V, Ghodiwala T.

OBJECTIVE

To analyse the results of the surgical treatment of patients with post-traumatic syringomyelia and to determine the optimal treatment tactics for patients with this pathology.

METHODS

During the period from 2010 to 2016 the authors treated 44 patients with post-traumatic syringomyelia, out of which 28 patients underwent surgery. The age of the patients ranged from 15 to 58 years. All patients underwent neuroimaging and were examined before and after the operation and were followed-up for a maximum of 54 months.

RESULTS

Patients were divided into 2 groups, 16 patients with uncomplicated spinal cord trauma and 12 patients with complicated spinal cord trauma. In the group of operated patients after uncomplicated spinal cord trauma, there was improvement in condition in 10 patients: 5 patients had no worsening of their condition and 1 patient had progression of syringomyelia. In the group of patients after complicated spinal cord trauma, 4 showed improvement in the condition, another 6 had no worsening and 2 patients had progression of syringomyelia.

CONCLUSIONS

The aim of surgery is to eliminate all factors that lead to obstruction of CSF flow. Shunting of a syrinx is a palliative operation with a high risk of recurrence. At the same time, arachnolysis with shunting can improve the results of treatment by reducing the volume of the syrinx, producing a rapid clinical effect. Using the chosen concept of treatment of post-traumatic syringomyelia in our group of patients allowed us to achieve stabilization or improvement in 9 out of 10 patients.

F16 CLINICAL STUDY OF SECTION OF THE FILUM TERMINALE IN THE TREATMENT OF TETHERED SPINAL CORD WITH SYRINGOMYELIA

Lou Y, Li H, Jin Y Liu L.

OBJECTIVE

The aim of this study was to assess the effectiveness of section of the filum terminale, intradurally and extradurally, in the treatment of syringomyelia due to tethered spinal cord.

METHODS

30 patients with tethered spinal cord and syringomyelia (15 males and 15 females) underwent minimally invasive surgery with section of the filum and expansion duraplasty. The pre-operative symptoms were related to lower limb sensory and motor nerve dysfunction.

RESULTS

Patients improved to different extents after release of the tethered cord. There was a statistically significant difference in the severity of symptoms at 1 year and at 3 years after the surgery. The syringomyelia also improved.

CONCLUSION

Section of both the intradural and extradural part of the filum terminale achieved good results in patients with tethered spinal cord and syringomyelia.

F17 PITFALLS IN CHIARI MALFORMATION TYPE 1 TREATMENT - POSSIBLE CAUSES OF SURGICAL FAILURES

Furlanetto M, Erbetta A, Saletti V, Chiapparini L, Babini M, Valentini L.

INTRODUCTION

There are many discussions about surgical indications and techniques for Chiari I malformation (CM) but few doubt that, when there is an associated symptomatic or evolving syringomyelia, craniocervical decompression with duraplasty is indicated. Failures of this first choice surgery are often complicated by progressive symptoms and slowly progressive neurological deterioration. The present study concerns 39 cases of revision Chiari surgery performed at the National Neurological Institute of Milan, between 1986 and 2018, 16 originally operated upon in our institution and 23 originally undergoing surgery elsewhere. In total they represent less than 10% of all cases (>350) initially operated upon at Besta in the same period.

METHODS

Surgical treatment was based upon the "failure pathogenesis" subgroups, which we classified into: a) uncomplete/wrong bone decompression (6 cases); b) unperformed duraplasty (11); c) CSF leak with "compressive" collection (4); d) excessive bone opening with "CBL sinking" (1); e) syrinx fenestration and intracranial hypotension (1); f) arachnoiditis, spontaneous or after untreated CSF collection (3); g) craniocervical joint instability (2); h) lack of resection of very low lying tonsils (4); i) inappropriate first surgery (4); j) unrecognized associated malformation (3).

RESULTS

One common finding was the high percentage (>50%) of patients requiring shunting/endoscopy for treatment of an associated hydrocephalus. Compared with our first surgery series, there were some cases of surgical morbidity and there was one case of post-operative mortality, caused by abdominal complications in an already highly compromised patient. Despite a good response of the syrinx to surgery, the neurological recovery was slow and uncomplete in the great majority of cases.

CONCLUSIONS

The results of "second look" operations can be good, if aimed at treating a recognized cause of failure. Often, permanent disturbance of CSF circulation is seen, caused by foramen magnum arachnoiditis and often needing treatment by endoscopy or shunting.

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F18 PREVENTING CSF LEAKS USING A PERICRANIAL GRAFT DURING CHIARI DECOMPRESSION Gopinath S.

ABSTRACT

In adults, Chiari decompression commonly involves duraplasty using synthetic materials. A significant proportion of those cases present later with CSF leak or pseudomeningocele. For the past several years, we have been using a pericranial graft to create a duraplasty and have not come across any patient with complications. We will describe our experience with a short video showing the technique.

F19 POSTERIOR CALVARIAL DISTRACTION FOR CHILDREN WITH COMPLEX CRANIOSYNOSTOSIS, CHIARI MALFORMATION TYPE 1 AND SYRINGOMYELIA - A 10 YEAR EXPERIENCE IN A UK SPECIALIST CENTRE

Lo WB, Thant KZ, Kaderbhai J, Rodrigues D.

INTRODUCTION

One-fifth of children with multi-suture and lambdoid synostosis have Chiari malformation type I (CM1). Posterior calvarial distraction is an effective method for increasing the intracranial volume in children with craniosynostosis. This study investigated the efficacy of this manoeuvre in posterior fossa volume expansion and treatment of CM1 with associated syringomyelia.

METHOD

A ten-year retrospective study in a quaternary unit.

RESULTS

Sixteen children were identified, 8 males and 8 females, with a mean age of 5.1 years, range 8 months to 19 years. Fourteen children had pan-synostosis and two had lambdoid synostosis. Eight were syndromic. Ten patients had raised intracranial pressure. Four had a syrinx. Clinically, 9 patients improved and 7 remained stable. None deteriorated. The average distraction distance was 23mm (range 16-28mm). An osteotomy extending inferiorly to the torcula was associated with a larger posterior fossa antero-posterior distance increase (13mm vs 6mm, $p=0.028$). The average tonsillar descent improved from 9.3 to 6.0mm. Syrinx dimensions also improved, the superior-inferior extent decreasing from 203mm to 136mm and the anterior-posterior diameter reducing from 7.9mm to 3.1mm.

CONCLUSIONS

Cranial volume expansion following posterior calvarial distraction also includes posterior fossa volume expansion. This results in improvement of tonsillar descent and syrinx. Posterior calvarial distraction is a safe and effective first-line treatment for children with concurrent multi-suture craniosynostosis, CM1, and syringomyelia. These findings also add to existing hypotheses that Chiari malformation is not a condition as such but a manifestation of an insufficient intracranial volume.

F20 SURGICAL TREATMENT OF PAEDIATRIC CHIARI I MALFORMATION - FORAMEN MAGNUM DECOMPRESSION WITH DURA LEFT OPEN, IN 70 PATIENTS

Kurzbuch AR, Jayamohan J, Magdum S.

INTRODUCTION

Surgery for symptomatic paediatric patients in our unit has consisted of limited sub-occipital craniectomy, C1 laminectomy, durotomy and arachnoid dissection, without dural repair. We carried out an audit of our results, to date, using this approach.

METHODS

A retrospective single-centre study, of 70 consecutive paediatric patients with Chiari I malformation, operated by two surgeons.

RESULTS

The mean age of this cohort of patients was 10.4 years, the youngest child being aged 2 and the oldest patient being 19. Thirty-two patients had syringomyelia as well as Chiari. Nineteen patients had concomitant scoliosis. Fifty-six patients (4 out of 5) reported post-operative improvement, 11 were clinically unchanged, and 4 noticed worsening of symptoms. Complications included CSF leaks in 6 patients, aseptic meningitis in 3 and CSF collections in 2 cases. Revision surgery was performed in 7 (1 in 10) patients. There was no long term surgical morbidity or mortality.

CONCLUSIONS

Foramen magnum decompression, with the dura left open, is a safe and effective surgical treatment in paediatric patients with Chiari I malformation.

F21 CHIARI I MALFORMATION WITH SIGNIFICANT MOTOR AND AUTONOMIC DYSFUNCTION IN A TODDLER

Naushahi M, Yates C, Sheen J, Ma N, Chaseling R.

ABSTRACT

Quadriparesis and cardiovascular instability are not symptoms commonly associated with Chiari I malformation diagnoses, less so in a 3-month-old toddler. Chiari I malformations have a wide array of symptoms, varying with regard to both the nature of onset and the age of the patient presenting. This variation is illustrated in this report, presenting a novel description of a set of symptoms in a particularly young patient, a case lacking the common insidious nature of Chiari I malformation symptoms. Acute onset of cardiovascular instability, global hypertonia, hyperreflexia, proximal upper limb weakness in a 3-month-old toddler are discussed. Parallels between the presented case and previous case reports are few, however some key features are distinguishable among similar cases. A successful posterior fossa craniectomy allowed the acutely unwell toddler to become a healthy, fully functional child. We believe this case contributes to elucidating the rare presentations of a curious pathology.

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F22 CHIARI I MALFORMATION – SHOULD WE OPERATE ON PICTURES OR CHILDREN? PROPOSAL OF A DIAGNOSTIC AND THERAPEUTIC FLOW CHART BASED ON THE RETROSPECTIVE ANALYSIS OF 630 MONO-INSTITUTIONAL CASES

Valentini L, Saletti V, Chiapparini L, Babini M, Furlanetto M.

INTRODUCTION

Many discussions about treatment for Chiari I Malformation (CM1) and syringomyelia in children centre both on indications and surgical technique. Complex Chiari is reported to need craniovertebral stabilization in as many as 50% of cases. The present review aimed to evaluate the results of craniovertebral decompression, with or without duraplasty and/or tonsillar resection, in a series of 150 operated children, focusing on the association with tethered cord and craniovertebral instability.

METHODS

171 children were operated for CM at the National Neurological Institute of Milan (FNCB) between 1986 and 2018. Their ages at surgery ranged from 1 up to 17 years. Preoperative MR was performed in every case and included the whole neuraxis, to rule out associated malformations. In 70% of cases symptoms before surgery were due to associated syringomyelia. A group of 459 symptomatic children were also followed, for a mean time of 5 years.

RESULTS

Only 20 asymptomatic children required surgery, despite pronounced tonsillar descent being present in many cases. In 22 cases upward migration of the tonsils was seen to occur over time. In the surgical group, pre-operative symptoms related to CM1 improved. Syringomyelia reduced in more than 80% of children and disappeared in a significant number. There was no major surgical morbidity and no mortality. Often, however, re-operation for tonsil resection was needed. Association with tethered cord was rare (2%). Craniovertebral junction malformations submitted to dynamic CT or MRI displayed mild instability in few patients but none has required fixation.

CONCLUSIONS

In children clinical symptoms are often more serious than in adults but surgical results may be better. True clinical and MRI instability requiring fixation is very rare. Asymptomatic children have a low risk of going on to develop symptoms.

F23 MAGNETIC RESONANCE IMAGING QUANTIFICATION OF MORPHOLOGICAL PARAMETERS IN CHIARI MALFORMATION – A RETROSPECTIVE STUDY

Fuell W, Elwy R, Albert G.

INTRODUCTION

Due to the significant presence of asymptomatic patients with Chiari I malformations (CMI) seen in paediatric neurosurgery, investigation of alternative morphological parameters in relation to symptomology could lead to revision of current diagnostic criteria. It is almost universally accepted that the caudal descent of the cerebellar tonsils from the posterior fossa into the upper cervical canal, of at least 5mm below the McRae line, constitutes a diagnosis of CMI. Although tonsillar herniation may aid in guiding diagnosis, tissue density within the foramen magnum and the role of posterior fossa volume development in relation to symptomology has not been explored from a diagnostic perspective.

METHODS

To provide a reduction in asymptomatic cases of CMI followed regularly, development and clinical implementation of a new diagnostic model based on the retrospective, longitudinal review of CMI patients was conducted. Tonsillar herniation, tissue density within the foramen magnum and posterior fossa volume were compared to signs and symptoms presented at each visit, to establish which measurement is found to be more tightly correlated.

RESULTS

Symptomology likely stems from the subarachnoid occlusion of cerebral spinal fluid at the foramen magnum, by the cerebellar tonsils, therefore making the tissue density within the foramen magnum a preferential candidate for assessment. Cases of patients outgrowing their CMI have been observed within the literature, which leads us to believe that, with an exponential growth in the posterior fossa, CMI resolution can occur, owing to the alleviation of hindbrain compression.

CONCLUSION

It is expected that tissue density within the foramen magnum is more correlated to symptomology than tonsillar herniation and differential growth of the posterior fossa could result in CMI resolution. With the current criterion for CMI, a considerable fraction of CMI diagnoses are incidental findings that would have previously not required follow-up, due to lack of symptoms.

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F24 HINDBRAIN HERNIATION IN HYPERMOBILE EHLERS-DANLOS SYNDROME PATIENTS

Smith FW.

BACKGROUND

A large proportion of patients suffering from the hypermobile Ehlers-Danlos syndrome (EDS) also show an element of hindbrain herniation, often labelled as a Chiari malformation.

METHODS

120 consecutive patients with hypermobile EDS had MRI scans of the cervical spine and craniocervical junction, performed in an upright MRI scanner, where the cervical spine angle was measured with the cervical spine in neutral, flexion and extension and compared with a group of normal individuals. Any instability of the cervical spine was recorded. The presence or absence of the atlanto-axial instability was also recorded. The presence or absence of hindbrain herniation was also recorded.

RESULTS

The majority of patients (80%) showed an increase in the cervical spine angle in neutral, which was 20% greater than that seen in the normal group. Fifty percent showed a greater ability to flex their neck and 45% had a greater angle in extension. Instability of the cervical spine was seen in 72% and 80% showed an element of instability of the atlanto axial joint. Evidence of cerebellar tonsillar ectopia greater than 5 mm through the foramen magnum was noted in 43%, with no evidence of syringomyelia. The pattern of hindbrain herniation was distinctly different from that seen in patients suffering from the Chiari syndrome. In the patients suffering from EDS, the herniated tonsils appear more rounded and are more laterally situated within the foramen magnum, when compared to patients with true Chiari syndrome, where the herniated tonsils lie more medial and are pointed in shape. Evidence of hindbrain herniation is seldom seen in the midline sagittal sections of an MRI scan.

CONCLUSIONS

With hindbrain herniation being more frequently seen in patients with EDS, it is important that the pattern of herniation is described differently from the Chiari syndrome, to avoid confusing the two different conditions.

F25 NEUROPHYSIOLOGICAL AND NEURORADIOLOGICAL CORRELATES IN PATIENTS WITH SYRINGOMYELIA AND CHIARI MALFORMATION - A CENTRAL MOTOR CONDUCTION TIME ALONG THE PHRENIC NERVE AND FIBRE TRACKING STUDY.

Ciaramitaro P, Massaro F, Ferraris M, Valentini CM, Cocito D.

INTRODUCTION

The cortico-diaphragmatic pathway was investigated by means of transcranial magnetic stimulation (TMS) in cases of amyotrophic lateral sclerosis (ALS) without clinical signs of respiratory impairment and was found to be a sensitive measure to reveal sub-clinical diaphragmatic impairment. This study aimed to: 1) investigate the central motor conduction time (CMCT) along the phrenic nerve in patients affected by syringomyelia, with or without Chiari Malformation (CM), in order to identify sub-clinical alterations of the phrenic pathway, and 2) to correlate the electrophysiological data to a Fibre Tracking (FT)-Diffusion Tensor Imaging (DTI) neuroradiological technique for evaluating cortico-spinal fibre loss.

METHODS

100 patients (25 males, 75 females), average age 49 ± 14 years, were selected and divided into three groups: 1) syringomyelia with Chiari malformation, 2) isolated syringomyelia and 3) isolated Chiari. TMS was performed in all patients by standard techniques. Electrophysiological parameters were statistically analysed and compared with 30 healthy subjects. DTI study was performed in 40 patients (altered/normal CMCT) and Fractional Anisotropy (FA) was calculated in three volumes of interest - whole cord, right and left hemi cord - at C2, C3, C4 spinal levels.

RESULTS

Pathological phrenic CMCT was observed in 27 patients (30% in subgroups syringomyelia + Chiari and isolated syringomyelia). FA values were significantly different at all levels, compared with healthy subjects (at C2 $p < 0.005$; at C3 $p < 0.05$, at C4 $p < 0.05$) and compared with altered CMCT patients at C3 level ($p < 0.05$), with a high correlation (76%).

DISCUSSION

CMCT along the phrenic nerve was a sensitive measure to identify alterations in voluntary respiratory pattern, before clinical presentation, in syringomyelia. Our results confirm the presence of an axonal damage underlying pathological CMCT.

CONCLUSIONS

Altered phrenic CMCT may be a good predictor of sub-clinical diaphragmatic impairment. The alterations found may be explained by cortico-spinal drive impairment and/or degeneration of the phrenic-related second motor neurons.

FREE PAPER ABSTRACTS

F26 IS CEREBROSPINAL FLUID PLEOCYTOSIS A GOOD INDICATOR OF POST-OPERATIVE ASEPTIC MENINGITIS FOLLOWING CRANIOVERTEBRAL DECOMPRESSION?

Asha MJ, Flint G.

OBJECTIVES

Differentiation between chemical and bacterial meningitis, following intracranial surgery, can be challenging. Current recommendations advocate use of broad-spectrum antibiotics in the presence of CSF pleocytosis, pending final cultures. We examined the implications of this recommendation in the context of craniocervical decompression for Chiari 1 malformation.

METHODS

Records of 225 patients with symptomatic Chiari 1 malformation, with or without syringomyelia, who underwent craniocervical decompression between 1995 and 2016, were reviewed retrospectively. Patients with clinically suspected meningitis were studied in detail. Initial CSF analysis was examined and results correlated with final microbial cultures. Other variables studied were patient demographics, presence of hydrocephalus, occurrence of postoperative CSF leak, use of prophylactic antimicrobials, length of hospital stay and clinical outcome.

RESULTS

34 of the 225 patients (15%) were suspected of having meningitis. CSF analysis showed pleocytosis in all these cases. The total white cell count was, however, widely variable and polymorphonuclear predominance was detected in 65% of cases. Final cultures confirmed that 3 of these 34 cases (less than 1 in 10) had bacterial meningitis. There was no significant difference in total white cell count, polymorphonuclear predominance, or CSF lactate between cases of aseptic and bacterial meningitis. CSF leak was documented in all 3 cases with bacterial meningitis but only two of the 31 patients with chemical meningitis. No difference in the incidence of hydrocephalus was detected between the two groups. Despite negative cultures, antibiotics were continued in half of all suspected cases where treatment had been commenced.

CONCLUSION

In the context of clinically suspected meningitis in the postoperative period, CSF pleocytosis is not a strong indicator for starting antibiotic therapy. Patients who appear clinically septic and/or have developed CSF leakage appear to be at higher risk of bacterial meningitis and should be considered for preliminary antibiotic treatment, pending culture results.

F27 CHIARI TYPE 1 MALFORMATION RELATED BLACKOUTS

Elhabal A, Flint G.

OBJECTIVE

Chiari type 1 malformation commonly presents with Valsalva-related headaches but blackouts also sometimes form part of the presenting profile. The objective of our study was to assess the frequency of blackouts and the effect of craniocervical decompression upon this symptom.

METHODS

A retrospective cohort study of Chiari type 1 malformation patients, with and without syringomyelia, who underwent craniocervical decompression between 2000 and 2016. We collected data on demographics and clinical symptoms of those patients whose symptomatology included blackouts.

RESULTS

We reviewed 241 patients operated upon for Chiari type 1 malformations between 2000 and 2016. We identified 161 patients who had only Chiari type 1 (group 1) and 80 patients with both Chiari type 1 and syringomyelia (group 2). Within the first group there were 16 patients (1 in 10) whose presenting symptom profile included blackouts. These blackouts ceased after surgery in all but one patient, in the follow up period. In group 2 we did not identify any patients who listed blackouts amongst their presenting symptoms.

CONCLUSION

Nearly all Chiari type 1 patients complaining of blackouts, as part of their symptom profile, enjoyed resolution of this symptom after decompression surgery, independently of any change in their other symptoms. The mechanism underlying Chiari-related blackouts warrants further discussion, particularly in view of their apparent lack of occurrence in patients with syringomyelia.

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F28 PRESENTING SYMPTOMS OF CHIARI MALFORMATION AS SEEN FROM THE PATIENT'S PERSPECTIVE

Burton L, Brereton G, Brereton N, Flint G.

INTRODUCTION

The Ann Conroy Trust support helpline receives an average of 140 calls each month. Callers are understandably concerned about the implications of their (usually) recent diagnosis of Chiari and/or syringomyelia. Not uncommonly, frustrations are aired about delays in diagnosis and an apparent unwillingness of medical professionals to accept the relevance of many presenting symptoms. Following on from a pilot study, we carried out a more detailed survey of patient experiences when being advised by medical professionals.

METHODS

A Survey Monkey questionnaire was constructed for patients diagnosed with Chiari malformation. Invitations to complete this were extended to potential participants via social media platforms used by patient support groups in a number of countries.

RESULTS

A total of 741 responses were received. Principal reported symptoms included headaches (88%), neck pain (75%), dizziness (68%), balance problems (67%), tingling/numbness in arms and hands (53%) and hearing disturbances (50%). Other reported symptoms included nausea and limb weaknesses. Whilst 37% of respondents were diagnosed within 6 months of seeking medical help, for 18% diagnosis took 6 years or longer. More than half (59%) did not feel that the specialist who diagnosed their condition understood it, with 48% being told that their Chiari malformation was an incidental finding. Following surgery Valsalva headaches resolved in 52% of cases. Other symptoms improved significantly in 38% of respondents, with 44% reporting a slight improvement. The remaining 18% reported no improvement.

CONCLUSIONS

Medical professionals vary in their ability to provide patients with acceptable explanations about their condition. Many patients therefore rely upon educational material prepared by dedicated support groups. Such information should never be taken as medical advice but can serve to help a patient better understand what a physician or surgeon is telling them during medical consultations.

F29 THE MECHANICAL ROLE OF OEDEMA IN CAVITY FORMATION

Venton J, Harris P, Phillips G, Hardwidge C.

INTRODUCTION

Oedema, sometimes referred to as the pre-syrinx state, has been noted prior to cavity formation, both clinically and in experimental syringomyelia. How this might contribute to cavity formation from a mechanical perspective is uncertain.

METHOD

A computer simulation of a two-dimensional, cross-section of the spinal cord has been developed. The cord was modelled as a poro-elastic material, which allows both the tissue and interstitial fluid to be examined. A region of oedema was modelled as an area of increased fluid content and flow in the posterior white matter.

RESULTS

Mechanical stress in the cord cross-section, potentially an indicator of cord damage, was calculated using the model. The region of oedema was found to cause increased mechanical stress in that area of the spinal cord, suggesting that damage is more likely to occur.

CONCLUSIONS

Further simulations of oedema in the spinal cord would be beneficial to understand how the presence of oedema increases the risk of cavity formation. A better understanding of the relation between oedema position and syrinx risk would be beneficial in determining the likelihood of syrinx formation, or of an existing syrinx worsening.

F30 COMPUTER SIMULATION OF SYRINGOMYELIA IN BRACHYCEPHALIC DOGS

Cirovic S, Lloyd R, Rusbridge C.

INTRODUCTION

Syringomyelia is a disorder associated with chronic pain, sensory loss, and paralysis. Since the pathogenesis remains unknown surgical interventions aimed at managing this condition have limited success. The most common cause of syringomyelia in humans is Chiari type I malformation and in animals the analogous condition is the Chiari-like malformation, to which some toy breed dogs are particularly predisposed. In cavalier King Charles spaniels up to 70% of dogs also develop Chiari-related syringomyelia. Such breeds thus represent a naturally occurring animal model to investigate the origins of syringomyelia. A finite element model of the canine spinal cavity has been developed to investigate the dynamics of spinal CSF compartment in normal and pathological conditions.

METHODS

The model included the spinal cord (with or without the syrinx) and cerebrospinal fluid (CSF) in the subarachnoid space, the dura and the epidural space. These features were reconstructed from MR scans of a cavalier King Charles spaniels suffering from a large syrinx. The cranial CSF compartment was modelled as a lumped compartment obeying the Monro-Kellie doctrine. The model was excited at the cranium to simulate shift in the blood volume related to the cardiac cycle.

RESULTS

For normal conditions the stress in the spinal cord was low and uniformly distributed along its length. With a blockage between the cranial and spinal CSF spaces normal shear and axial stresses increased significantly at the regions in the cord where the syrinxes typically form. This elevated stress originated from the bending of the cord at the locations where its curvature was high.

CONCLUSIONS

The results suggest that repetitive stressing of the spinal cord, caused by its exaggerated movement, could be a cause for the initial formation of syrinx cavities. A more comprehensive model of cranial CSF compartment and consideration of factors such as cord tethering is needed.

FREE PAPER ABSTRACTS

F31 SYRINGOMYELIA PATHOLOGY - INSIGHTS FROM ANIMAL MODELS AND ULTRASTRUCTURAL STUDIES

Lam M, Hemley S, Najafi E, Berliner J, Bilston L, Stoodley M.

INTRODUCTION

Syringomyelia includes a range of conditions characterized by the presence of fluid-filled cavities in the spinal cord. The mechanisms of syrinx formation remain poorly understood and treatment options are limited. Perivascular spaces have been hypothesized to play a role by providing rapid access for cerebrospinal fluid in the subarachnoid space into the spinal cord tissue but their anatomical details have not been well described.

METHODS

We used transmission electron microscopy to study the ultrastructure of perivascular spaces and the surrounding spinal cord tissue, in healthy Sprague Dawley rats and in a model of post-traumatic syringomyelia. The model was created using a motorized spinal cord impactor, followed by a subarachnoid injection of kaolin to produce arachnoiditis. In healthy animals, CSF tracers (horseradish peroxidase, ovalbumin-AF647; 45kD, and nano-gold, 5 nm) were used to investigate the continuity of fluid pathways between subarachnoid space and central canal.

RESULTS

Animals with post-traumatic syringomyelia appeared to have an intact blood-brain barrier. However, spinal cord perivascular spaces were strikingly enlarged. Perivascular spaces were continuous with the extracellular spaces of the surrounding tissue and with the vascular wall basement membranes. Other abnormalities included an abundance of immune-type cells in the subarachnoid space, broadening of extracellular spaces and loss of tissue integrity. Five minutes after injection at cisterna magna, CSF tracers were distributed throughout the tissue and fluid compartments. Surprisingly, tracers were also seen in the lumen of blood vessels, suggesting trans-vascular clearance.

CONCLUSIONS

These findings have potential implications for CNS fluid volume regulation, as well as clinically for the detection of CNS-derived biomarkers in blood, the regulation of immune response and for delivery of therapeutic agents.

F32 HISTOLOGICAL ANALYSIS OF ARACHNOID FEATURES IN CHIARI I MALFORMATION

Klekamp J, Heidary M.

INTRODUCTION

Although foramen magnum decompression is accepted as the treatment of choice for Chiari I malformation, the surgical technique is controversial. Do we need to open and dissect the arachnoid? Do we need duraplasties? This paper focusses on histological features of the arachnoid in these patients.

METHODS

162 consecutive arachnoid samples, taken routinely during foramen magnum procedures over a period of 10 years, were analysed, measuring thickness, cellularity and the amount of fibrosis. These measurements and evaluations were performed blinded for any clinical data or intra-operative findings. A classification into three grades of arachnoid features was created. These histological grades were compared with intraoperative evaluations of arachnoid changes and their relevance for pre-operative symptoms and post-operative results was analysed.

RESULTS

Arachnoid features were found to be quite variable in terms of thickness, cellularity and the amount of fibrosis, each following a normal distribution. The histological grades correlated significantly with intraoperative evaluations of the arachnoid, as noted by the surgeon after dural opening. Highly significant correlations were found between histological grades and the severity of pre-operative gait problems, motor weakness and sensory disturbances. On the other hand, intraoperative evaluations of arachnoid changes by the surgeon showed significant correlations with the presence of syringomyelia, the postoperative clinical result after 3 months and rates for progression-free survival in the long-term.

CONCLUSION

Although this study cannot prove a causal relationship between arachnoid features and clinical symptoms or postoperative results, it clearly shows that the arachnoid should not be considered irrelevant in patients with Chiari I malformation.

F33 POSTERIOR FOSSA VOLUME OF CHIARI MALFORMATION - IS IT FUNNEL NARROWING?

Liu Y, Li H, Jin Y, Liu L.

OBJECTIVE

To study the pathogenesis of Chiari I malformation with volumetric measurements of the posterior fossa.

METHODS

We compared the posterior fossa volume of 59 patients with Chiari I malformation with a control group from the normal population. The volume was measured on MRI scans with the use of Mimic software.

RESULTS

The ratio of PFB to PFC of each image layer increases from the rostral end to the caudal end in the patient group while it decreases in the control group.

CONCLUSIONS

We presume that Chiari I malformation is caused by the narrowing of the lower part of the posterior fossa, i.e. a funnel narrowing. Clinically, it is impractical to improve the ratio with an approach involving focal surgery. Instead of arguing which surgical approach is optimal, we should concentrate on restoring/improving the CSF circulation with surgical techniques. A minimally invasive technique that can improve the CSF circulation should be customized for each patient individually.

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F34 ASSESSING HUMAN BRAIN TISSUE SLICES DERIVED FROM CEREBELLAR TONSILLAR TISSUE IN CHIARI PATIENTS, AS A LABORATORY BASED INJURY MODEL.

Sen J, Tickle JA, Adams C, Price RF, Harrisson S, Tzerakis N, Chari DM.

OBJECTIVES

Use of animals to study CNS repair is controversial. Concerns relate to the potential for animal suffering and failure of animal models to predict human responses. This study aims to address these issues by attempting to create a reliable dish model of brain injury using human brain tissue.

METHODS

Neuroscientists and neurosurgeons collaborated to grow slices of brain removed from Chiari malformation patients during decompressive surgery. Adult patients with Chiari malformation consented to take part in the investigation. In consented patients in whom it was deemed clinically beneficial to excise cerebellar tonsil tissue as part of the procedure, tissue was transferred directly to the laboratory for processing. Viability assessment and tissue responses to injury were investigated and compared with known in-vivo tissue responses.

RESULTS

Two patients thus far have been enrolled. Derived slices were maintained alive or showed viability in growth medium and survived for beyond 40 days (live/dead stain). The major neural cell types were detected and remained viable over this time period.

CONCLUSIONS

Human brain slices can survive ex-vivo. Major neural cells can be detected. Focal trauma elicited a stereotypical response. This model could have a significant impact in reducing animal usage and offer potential progress to testing novel therapeutic / neuroprotective interventions and also possibly ensure rapid progression of effective and safe therapies for CNS disorders, improving reliability and patient safety.

F35 MANAGEMENT OF PREGNANCY AND DELIVERY IN WOMEN WITH CHIARI MALFORMATION TYPE I AND/OR SYRINGOMYELIA - A VARIABILITY SURVEY

Knafo S, Picard B, Samadi M, Benhamou D, Parker F.

ABSTRACT

Management of Chiari malformation type I (CMI) and syringomyelia during pregnancy and delivery remains debatable. The aim of this study was to identify current practices and to investigate whether some of these may have an impact on the natural history of the disease.

METHODS

A retrospective cohort study of women harbouring CMI and/or syringomyelia, born between 1970 and 2000, and followed by the French National Centre for Chiari and Syringomyelia, in Bicêtre Hospital. Among 311 patients eligible, 99 women with at least one delivery were included in the final analysis.

RESULTS

Seventy percent of patients had two or more deliveries. Forty-one percent had surgery for either CMI and/or syringomyelia. Among these 71% had surgery after their first pregnancy. Sixty-five percent of patients underwent vaginal delivery, 19% had a scheduled Caesarean procedure and 16% underwent emergency section. Only 21% of patients declared that the delivery method was chosen in consultation with their neurosurgeon but amongst these the rate of scheduled Caesarean procedures was 52%. Fifty-nine percent of patients had epidural anaesthesia (38% after neurosurgical advice) and 19% had a general anaesthetic. There were no complications related to pregnancy in 79% of cases. During pregnancy, only 13% of the patients reported a worsening of their symptoms, 68% were unchanged and 10% were improved. After pregnancy, 34% of patients reported some worsening of their symptoms but only 3 patients were diagnosed in the year following their pregnancy. There was no difference regarding the proportion of clinical deterioration between the modalities of delivery or anaesthesia.

CONCLUSION

The majority of patients harbouring CMI and/or syringomyelia undergo vaginal delivery and epidural anaesthesia without complication. However, when solicited, most neurosurgeons still favour Caesarean procedures under general anaesthesia. A consensus between neurosurgeons, obstetricians and anaesthesiologists would be desirable, to prevent unnecessary Caesarean procedures.

F36 PREGNANCY AND LABOUR CARE FOR WOMEN WITH CHIARI MALFORMATION - A CASE SERIES

Roper JC, Al Wattar BH, Pirie AM, Silva A, Flint G.

INTRODUCTION

Care for pregnant women with Chiari malformation is heterogeneous, with concerns about raising intracranial pressure during labour leading to worse neurological outcomes and rising caesarean section rate. We examined care provision in a large tertiary centre to identify elements of best practice.

METHODS

A retrospective case series of all pregnant women diagnosed with Chiari malformation at the Birmingham Women's hospital, over the ten years between January 2004 and August 2014.

RESULTS Eleven women diagnosed with Chiari malformation were identified. Nine of those women had a planned normal vaginal delivery and two underwent elective Caesarean section for non-neurological reasons. There were no neurological complications recorded during pregnancy or delivery.

CONCLUSIONS

Uncomplicated Chiari malformation is not a contraindication for vaginal deliveries.

FREE PAPER ABSTRACTS

F37 MRI BIOMARKERS FOR CANINE CHIARI MALFORMATION-ASSOCIATED PAIN AND SYRINGOMYELIA

Spiteri M, Wells K, Knowler SP, Rusbridge C.

INTRODUCTION

Canine Chiari malformation (CM) is prevalent in brachycephalic toy breeds of dog, including the cavalier King Charles spaniel (CKCS). Although some dogs are asymptomatic, CM can be associated with pain and secondary syringomyelia (SM). Morphometric studies on traditional MR images can distinguish between clinical groups but it is not easy to translate findings from research into clinical practice. The aim of this study was to extract markers from MRI in relation to CM associated pain and SM in adult CKCS dogs.

METHODS

This study was split into two analyses: 1) comparing a symptomatic CM pain class to asymptomatic CM controls, and 2) comparing a symptomatic SM class to the same control group. Diagnosis was based on clinical signs and consented MRI. A midline sagittal MRI of the head and neck of a control group dog was chosen as a reference. Midline sagittal MR images of 77 dogs were mapped onto the reference MRI using DEMONS (non-linear) image registration, producing a 2D deformation map for each case. For each pixel, direction and magnitude of the mapping deformation were computed. Potential biomarkers were identified amongst these descriptors using a feature-selection algorithm, to identify candidate markers of CM pain or SM, and a kernelised Support Vector Machine classifier, to analyse the ability of these to separate controls and clinical cases.

RESULTS

The area under the curve was 81.51 for CM pain and 86.10 for SM. Analysis identified 5 markers for CM pain, in the regions of the nasopharynx, soft palate, caudal nucleus hypothalamus and 4th ventricle. It also identified 5 markers for SM, in the regions of soft and hard palate interface x 2, soft palate, trochlear nucleus, and corpus callosum.

CONCLUSIONS

Identification of biomarkers can be used to develop an objective tool for diagnosis.

F38 VESTIBULAR SIGNS, AUTONOMIC DYSFUNCTION AND DYSPHAGIA MIGHT OCCUR IN ADULT DOGS WITH SYRINGOBULBIA

Williamson B, Davies E, Epperly E, Roynard P, Scrivani PV.

INTRODUCTION

Syringobulbia is a pathologic condition characterized by the presence of one or more fluid-filled cavities within the brainstem. This retrospective case series describes clinical and radiological findings in eight dogs with syringobulbia.

METHOD

Syringobulbia was diagnosed during magnetic resonance imaging (MRI) carried out for evaluation of intracranial disease. Five dogs also underwent repeat MRI, at intervals ranging between 6 and 55 months. All eight dogs were adult, small-breed dogs with concurrent syringomyelia.

RESULTS

Six dogs had evidence of central vestibular disease on admission. On MRI, the fluid-filled cavities had signal intensity characteristics like cerebrospinal fluid, were in the medulla oblongata and were solitary in each dog. Initially, the shape of the cavity was a slit in five dogs and bulbous in two dogs. On repeat MRI, one dog had progression of syringobulbia from slit-like to bulbous. Four dogs remained as unchanged, slit-like syringobulbia. One dog had MRI prior to development of syringobulbia and, following cranioplasty, a slit-like syringobulbia was detected. A variety of medical and surgical treatments were performed with improvement of some but not all signs. One dog died following surgery due to cardiopulmonary failure and the other seven dogs were alive at least one year after the initial diagnosis, which was the minimal time of follow-up. One surviving dog developed a unilateral hypoglossal nerve deficit two months after the initial diagnosis and mega-oesophagus 14 months later.

CONCLUSION

Detecting a fluid-filled cavity in the medulla oblongata, consistent with syringobulbia, is possible in dogs undergoing brain MRI. The cavity may be slit-like or bulbous, progress or remain static, and might be associated with syringomyelia, vestibular signs, autonomic dysfunction, and dysphagia.

F39 PERSISTENT FONTANELLES IN CHIHUAHUAS.

Kiviranta A, Rusbridge C, Lappalainen AK, Vapaavuori O, Jokinen TS.

INTRODUCTION

Anecdotally, a persistent fontanelle is common in adult Chihuahuas but the prevalence is unknown. Known as a molera, or "soft spot", it is considered a mark of "purity" by some breeders. The aim of this study was to describe the presence and distribution of persistent fontanelles in Finnish Chihuahuas, as part of a broader study investigating Chihuahuas with Chiari-like malformation and syringomyelia.

METHODS

Data was collected from both symptomatic dogs and those without clinical signs. The cranium of the dogs was imaged with computed tomography. Dorsal, lateral and caudal cranial sutures were assessed for the presence of persistent fontanelles.

RESULTS

Fifty dogs were included into the study. Forty-six (9 out of 10) had a persistent fontanelle. Thirty-three (2 out of 3) had between 1 and 3 persistent fontanelles but with a range in the whole group of 1 to 13. Most of the persistent fontanelles were located dorsally (58/250, or 1 in 4), and most commonly at the fronto-parietal suture (40/50 dogs, or 4 out of 5).

CONCLUSIONS

No previous reports describing persistent fontanelles in dogs exist and the clinical significance of these is currently unknown. In children, persistent fontanelles are associated with increased intracranial pressure. Our future aim is to evaluate possible association of persistent fontanelles with the presence of syringomyelia, ventriculomegaly and mastoid foramen areas, as well as with Chiari-like malformation and syringomyelia-related clinical signs.

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