LMBBS Conference Report 2010

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Introduction

The LMBBS Annual Family Conference was, once again, a weekend packed with activities, fun and laughter, where old and new members met to share experiences and learn about the latest research within the wonderful Hilton setting.

So successful has this weekend become that members and families are arriving earlier each year, and are starting to ask, 'Please can we come Thursday to Monday' and the children want to know why they can't have two Theme Park Trips! (Not sure that the Carer's could cope with this!!)

The success of the weekend is down to ‘team work’ and organization and Conference 2011 is already in the planning stages. Your evaluation reports are invaluable and are used to ensure that any problems or suggestions are dealt with before the following Conference. Your advice and requests regarding Professionals and speakers are again important; this is your Conference and we want to get it right.

A question often asked is ‘Why choose Northampton as a venue?’ Apart from the fact that the food and hospitality is excellent and the hotel is centrally located for travelling by bus, air, rail and car, it is the Collingtree Hilton Hotel and staff who have, over the past 10 years, improved and excelled in their precision planning for this event, especially with regards to Disability Awareness. Their attentiveness to our guests, especially to those with a visual impairment, is second to none. VI guests are guided to their tables and a personal waiter/waitress is available to plate meals and ensure that guide dogs have a plentiful supply of fresh water. It is heart warming to be greeted annually by the same Admin and Front House staff, who meet our needs so well.

The Conference wouldn’t be a success without the excellent mix of speakers and facilitators, who give up their precious weekend time so willingly, to report on the latest medical updates in LMBBS; these talks are all reproduced in this Conference Report. During the afternoon, the delegates have the opportunity to ask questions in a smaller setting and it is evident, from the comments of those who attend, that they would like the workshops to be longer; however time is limited. This year, to end the afternoon, we had a very moving perspective from Emma Turnbull.

Research is immensely important to all our members and the Conference Weekend often allows us to play our part. This year was no exception and, once again, our members excelled in signing up for the ‘sleep apnoea’ research programme, with Dr Mathew Hind and Dr Robyn Quinlan.

A big THANK YOU goes to the ‘unsung heroes’ of the Conference, our volunteer carers, who have been coerced over the years onto the team. They enable parents to enjoy the Conference, in the knowledge that their children/young adults are being expertly looked after, either in the creche, enjoying the many toys and craft activities, or visiting a Theme Park and hanging upside down on some of the rides. After only an hour’s break on their return, the carers are available once again to allow parents a child free evening meal, if they wish, and the opportunity to socialise. On a personal note, a big ‘Thank You’ to the Creche Team for taking such good care of my ‘little one’ which enabled me to concentrate on the Conference, knowing she was so well looked after. It is evident from the evaluation reports how grateful parents are for this opportunity.

Thanks go to Sue King, from Guide Dogs for the Blind, Gloucestershire, who has been coming to the Conference for at least ten years and works with the Care Team on guiding skills. She also raids the resource room at Guide Dogs for the Blind, filling her car with everything from a talking microwave and scales to clocks, watches, games and much more.

Sue has been known to miss the 'Cup Final' to attend, so, for this, we are eternally grateful! Many thanks also go to Ray Perry, who was on hand, once again, to provide benefits help and advice.

Two new exhibitors this year were Debi and Trevor Platt who brought along a selection of products for the visually impaired, including sunglasses. Many of our members took the opportunity to try out the Sonar Cane. LMBBS has been invited to attend their Sight Awareness weekend in Manchester, later in the year, which is unchartered territory for LMBBS; however, we are very much looking forward to the opportunity of forging new links.

As usual, there was a resource table in the hotel lobby, which, as well as displaying LMBBS material, provided useful information and contacts from other organisations for people to take home. The fundraising table had a tombola, which was great fun for all, especially the children. On the Saturday evening, there were various activities available, including pool tables, table top games, board games and a huge craft table. New Committee Member, Richard Zimbler, entertained the children/young adults with a karaoke and the Quiz Night and raffle ended the day on a high note for everyone.

As Trustees of the Society, we are totally committed to the annual organisation of the LMBBS Family Conference and planning for 2011 is already underway. We have two speakers flying in from the USA and Strasbourg and we have interested families from Spain and France. Nonetheless, it is you, the members and professionals, who actually make the Conference such a resounding success. Your continued support and appreciation of the weekend makes all the work worthwhile and we look forward to seeing you all again in 2011.

Chris Humphreys

Update on Research and Study of LMBBS

Professor Phil Beales
Professor of Medical and Molecular Genetics
Wellcome Trust Senior Research Fellow in Clinical Science,
Honorary Consultant in Clinical Genetics, Molecular Medicine Unit,
Institute of Child Health/Great Ormond Street Hospital for Children and Guys and St Thomas' NHS Trust,
President of LMBB Society

Professor Beales obtained his degrees in Genetics and Medicine form University College, London. He undertook postgraduate training in both general medicine and paediatrics before specialising in Clinical Genetics. In 1999 he published the largest survey of LMBBS problems with the help of members of the Society. This culminated in the identification of a number of new features not previously described in the medical literature and has helped Professor Beales to propose new diagnostic criteria with the expressed purposed of enabling an earlier diagnosis in many children.

In 1999, Professor Beales spent over a year at Baylor College of Medicine in Houston, where together with Nico Katsanis, Richard Lewis and Jim Lupski, they discovered the first gene (BBS6) to cause LMBBS. Since then, at least 12 genes are linked to the syndrome. Professor Beales now works jointly as a Consultant Clinical Geneticist at Guys Hospital, London and the Institute of Child Health, at Great Ormond Street Hospital, and runs a laboratory research group at the latter. Together with collaborators from Europe and North America, his group have made major advances in our understanding of the causes of the syndrome. This includes the notion that abnormally functioning cilia (small finger-like appendages on cells) lies at the heart of LMBBS. The challenges that lie ahead involve understanding how dysfunctioning cilia contribute to various syndrome aspects. These

discoveries bring closer the ultimate goal for the design of potential treatments to prevent further visual deterioration or weight gain.

Professor Beales has been medical advisor to the LMBB Society since 1996 and was made President of the Society in 2005. In 2010 he succeeded, with the help of the Society, in attracting funding from the DoH to establish national multidisciplinary clinics and a comprehensive genetic testing platform for all persons with LMBBS.

Professor Beales welcomed everyone and began by introducing the speakers and outlining the day's programme. He then proceeded to give a research round-up of what he and his team have been up to over the past twelve months, in particular, working on one of the BBS genes, BBS9, which Professor Beales thinks may be the 'Bridge of the Enterprise, the one that seems to hold it all together'. Professor Beales and Anna Diaz-Font, from his team, visited patients towards the end of 2009 in connection with their work on a further gene, which is thought to modify or affect the way that LMBBS might present, in terms of the age of onset of visual problems and so on. Professor Beales introduced Beth Hoskins, a member of his team and a familiar face at the family conference weekend, who has just completed a mutation screen and written a paper on the data obtained. Dan Osborn, Miriam Schmidts, Aoife Waters and Victor Hernandez were also introduced, and they are working on other research projects, including cilia biology, organ development, drug screens and so on.

Dan Osborn and Miriam Schmidts have been working on a drug library screen, and it is hoped that, at the next Conference or so, they may be able to present some of their work. The area they have been looking at in particular is kidney cysts. They have been working with fish that have been genetically engineered to have LMBBS and kidney cysts. Different drugs are added to their water and they observe the fish for any improvement in their condition. It takes ten to fifteen years to develop a drug from the minute it is discovered so the team are using an approach called 're-profiling', using medicines that are already licensed and in use for other conditions. Because these drugs are already safe to use in man, any that prove to be useful can quickly be trialled without delay. There have been a number of promising leads from this study; however, it is still early days.

Professor Beales briefly mentioned research concerning weight maintenance issues, which is another difficult aspect of LMBBS. In particular, they are looking at the pancreas and the potential connection between cilia, weight maintenance and sugar control and so on. There has also been research into immune function and lung function in LMBBS, following the study that was undertaken at last year's Conference; further details of that will follow.

Professor Beales then moved on to talk about the BBS Multidisciplinary Clinics which are now underway. He said that the purpose of the clinics is to provide a one-stop-shop so that patients can come on one day and have most, if not all, of the major components of the Syndrome looked at by a specialist. Professor Beales has been running a BBS clinic for over ten years, however, as he said, he is a geneticist, not a kidney specialist, ophthalmologist or endocrinologist, so it's better that all these specialists are brought together. Professor Beales said, 'It's been no mean feat. After about 800 or so emails, 200 letters and about another 300 documents, I can stand here and say we finally did it.'

Professor Beales reported that the service, which was up and running from the 1st April 2010, consists of five main clinical partners, spearheaded by the LMBB Society. The reason for choosing the centres was two-fold; they had to divide the country up reasonably fairly to ensure ease of access, and choose centres where the expertise already exists. Birmingham

became the natural centre where Birmingham Children's Hospital and University Hospital, Birmingham look after the north of the country, with Great Ormond Street Hospital and Guys Hospital, London looking after the south. Professor Beales also managed to convince the Department of Health of the need to have a new DNA laboratory and he was delighted to say that Beth Hoskins has been appointed to one of the Clinical Scientist posts to set up and run that particular service. To sum up, Professor Beales said that this is an outpatient-based service and is a multi-disciplinary clinic, involving an Ophthalmologist, an Endocrinologist (hormones), a Nephrologist (kidney specialist) and in some places a Psychologist, Geneticist and Dietitian. There will be a dedicated nurse, and, of course, overseeing all of this is the LMBB Society.

Professor Beales finished by talking everyone through a typical day at Great Ormond Street Hospital's BBS Clinic. On arrival, patients are met by the LMBBS representatives who will be on hand to provide help and support throughout the day. Patients are given a timetable, letting them know where they need to be and when. The morning is spent in the Ophthalmology department, under the care of Ophthalmologist, Will Moore, where very detailed visual assessments will be done, which is obviously important for children. The adult's clinics won't have quite so many tests. First, the peripheral vision is measured, then there is a test called an ERG, which assesses the function of the back of the eye to see how good it is at receiving light and receiving images, and getting this information back to the brain. Photographs are taken of the back of the eye to assess the thickness of the retina and also to see how the back of the fundus of the eye looks, and all of these tests will give an idea of exactly what's going on and what the state of the eye is.

At some point during the morning, there is a blood test to check kidney function, liver function, hormones, full blood count among other things, as well as a detailed genetic test. That all comes out of just the one blood test and there will be 'magic cream' or spray for those who need it. Lunchtime is spent next door in the Homoeopathic Building on Level 2, where the dietitian, Sarah Flack, will have laid out an array of sandwiches, vegetables and dips and a selection of fruit. For the afternoon session, patients are given a room and each clinician will visit for a 25 minute consultation. Wendy, the play worker, ensures there are age appropriate activities in each room to keep the children busy. Following the appointment, patients will receive a letter detailing all the results from the day, with a copy being sent to the local healthcare team for optimum ongoing care.

Professor Beales finished his talk by inviting questions from the audience and a young lad asked if more blood would be taken than usual for all the tests! Professor Beales reassured him that there would be plenty left behind! Further questions were:

How do you get a referral to the BBS multi-disciplinary clinics?

Professor Beales replied that all those on the LMBBS membership database would automatically receive an invitation sometime over the next 18 months. Others would be referred by their Specialist or GP.

I know there's an issue as far as people coming from different parts of the country. Could you clarify that?

Professor Beales replied that, although initially the Department of Health were excluding Wales, Chris Humphreys had been lobbying the Assembly and indications are that everyone from Wales will have access to the service, as will those from Scotland and Northern Ireland.

Renal Diseases in Adults

Dr Lukas Foggensteiner

Consultant Nephrologist, Queen Elizabeth Hospital, Birmingham

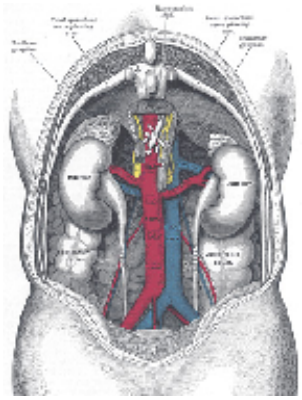
Dr Lukas Foggensteiner has been a consultant kidney specialist at the University Hospital, Birmingham since 2002 and, prior to that, was a trainee in Addenbrooke's Hospital, Cambridge. He has a continuing interest in genetic kidney diseases and did a PhD on the subject of polycystic kidney diseases in Cambridge University in the late 1990s. In 2004, he established a specialist clinic for genetic renal disease in conjunction with the clinical geneticist in Birmingham which provides diagnosis, treatment and counselling for a range of patients with inherited renal disease, including Bardet-Biedl syndrome. In April 2010, Birmingham, led by Dr Foggensteiner, became one of two national centres providing a specialist service for adults with LMBBS as part of an initiative funded by the National Commissioning Group. Dr Foggensteiner also continues to be involved in research projects, developing treatments for genetic renal disease, as well as working as a general nephrologist, undertaking dialysis and kidney transplantation in one of the largest regional renal centres in the UK.

Dr Foggensteiner began his talk with a brief outline of basic genetics. There are millions and millions of cells in the body and in every cell there is a nucleus. In every nucleus there are chromosomes and these are made up of very tightly coiled loops of what we call DNA. James Watson and Francis Crick, who were working out of a Lab at Cambridge, were frequent lunchtime patrons of the Eagle Pub and, at lunchtime on 28th February 1953, they announced they had "discovered the secret of life" after they came up with their proposal for the structure of DNA. Dr Foggensteiner joked that, although he had been to the Eagle pub many times, he'd not had any similar kind of inspiration.

The talk continued with an explanation of how mutations in DNA cause genetic disease. The DNA itself consists of huge numbers of base pairs, and these base pairs essentially form a kind of code, an alphabet, and there are three billion of these base pairs in every human genome. It helps to think about it in terms of a printing error in a very large book; it needs just one letter to be wrong in your entire volume of the Encyclopaedia Britannica to cause the problems that constitute genetic disease. The delegates were shown a picture of a Rolls Royce Merlin engine taken at Duxford War Museum and it was explained that the engine is the final product, much like the human body, and the plans to build the engine are like our genes. If there is a printing error in the plans and the size of one of the components is wrong, when the factory tries to manufacture that component, using the wrong dimensions, then the engine simply won't work, or, if it does work, it might not work properly and that's really how genetic disease happens. With BBS, the challenging and interesting thing is that there are lots of different genes that cause the condition, so, going back to the engine, there are different mistakes in different plans for different components that seem to result in the same outcome, which makes this genetic disease quite challenging to manage in many respects and is probably also why each individual with this condition is affected so differently. However, investigating genes and sequencing them has been made a lot easier by the human genome project, which has meant that all of our DNA has been sequenced and made available to scientists, certainly one of the great achievements of the last decade.

Dr Foggensteiner moved on to discuss Bardet-Biedl Syndrome in more detail, focusing briefly on the prevalence of the condition. He suggested that when considering how common a condition is, to imagine you're a GP and whether you would come across a case of this condition in your practice. A GP will typically have about 3000-5000 patients on their books, so will they see a case of BBS? Well, probably not because it is pretty rare; even specialist doctors in big teaching hospitals may only see very few cases and that is one of the reasons why a specialist clinic focused in a couple of hospitals around the country is a

very good idea. Dr Foggensteiner then moved on to discuss the phenotype of BBS, the symptoms of the syndrome, in particular how it affects the kidneys.



The delegates were shown an engraving taken from Gray's Anatomy; there are two kidneys, left and right, connected to the bladder via tubes called urethras. The kidneys clean the blood, filter out the toxic substances (the by-products of metabolism) which leave the body in the urine.

So what goes wrong in Bardet-Biedl Syndrome? Well, the abnormalities that we see in the kidneys in Bardet-Biedl Syndrome are variable. Often the shape of the kidneys is abnormal, the way that the kidneys have developed may not be entirely normal, and, if we look closely, there may be abnormalities in the kidneys of a very large proportion, if not all, of those with the syndrome. Although lots of people with BBS may have abnormally developed kidneys, the good news is that most of them will have normal kidney function. From the patient viewpoint, it doesn't matter what the kidneys look like, it is whether they work that the doctors are actually interested in and kidney failure is actually pretty uncommon in BBS. It is, however, a troublesome problem and it is important that declining kidney function is spotted early. Dr Foggensteiner emphasised the importance of checking blood pressure because high blood pressure can damage the kidneys and damaged kidneys, in turn, can cause high blood pressure.

It was then explained how kidney function is tested, which is through measuring creatinine levels in the blood. Creatinine reflects kidney function fairly well and, if your creatinine level is high, it may mean that your kidneys are not working particularly well. A further test is the estimated glomerular filtration rate, or eGFR, which is calculated from creatinine levels, sex and age and works best on average-shaped bodies. Unfortunately, the test is less useful for those who are short and overweight or tall and thin. It was suggested that a lot of doctors, even kidney doctors, seem to forget this and patients with BBS who may not be of average height and weight may need to remind their doctor of this, should the eGFR come back a little abnormal. Other tests that are used to check kidney function are ultrasound scans, CT scans and MRI scans.

Dr Foggensteiner moved on to talk about potential treatments and therapies. In polycystic kidney disease, there are some drugs that are going through clinical trials that may affect how quickly the cysts grow. It is not yet known whether this is relevant to LMBBS; however, there are more general things that affect the kidneys and how quickly kidney disease progresses, irrespective of what the cause is. As mentioned earlier, blood pressure is really important because high blood pressure will make any kidney disease of any type worse, and it is known that if you treat blood pressure, the disease slows down. Anaemia is a common complication of kidney disease and can be treated with EPO injections which increase the levels of haemoglobin in the body. Iron and Vitamin D tablets are often necessary as well, and dieticians may recommend special diets to avoid potassium and phosphate and so on. So, to help yourself, check your blood pressure every few months. You can buy a home testing device from Boots for around £20; however, it is important that the cuff fits properly, otherwise the reading will be inaccurate. Make sure you get your kidney function checked every year, and, if diabetic, make sure your diabetes is well controlled. Diabetes itself can cause kidney problems (diabetic nephropathy) and someone with BBS who is diabetic is just as vulnerable to diabetic nephropathy as any other diabetic, so controlling it is important. Weight control is also important because extra weight can put a strain on the kidneys and, if you do end up with kidney failure, it can be difficult to do a transplant on someone who has a lot of weight around their tummy. He continued, "It goes without saying that you shouldn't smoke and you should get plenty of exercise as well. All of these general health tips will

benefit your kidneys as well as your heart, lungs and the rest of you". Dr Foggensteiner said that he often gets asked about stem cell therapy and gene therapy, but, unfortunately, they are not yet applicable to those with genetic disease and, to date, they have been unable to grow kidneys, using such technology.

As mentioned previously, it is crucial that with BBS the kidneys are checked regularly because, with early renal disease, there are often no symptoms at all and most people with kidney disease don't know they've got it until their kidney function has got to the point where it's not good enough to keep them well anymore. Occasionally, the cysts can cause pain, usually around the back, or there may be blood in the urine. It was reiterated that renal failure doesn't happen in many people with Bardet-Biedl Syndrome; however, a minority will be affected. Renal failure can be treated with dialysis and it can also be treated with a kidney transplant. It was explained that dialysis is a treatment that was established in the 1950's and is now a routine method of replacing the function of the kidneys. There are two types of dialysis that are available; one is called peritoneal dialysis, or PD, the most common form, and the other is haemodialysis. The principle of peritoneal dialysis involves putting fluid into the tummy through a silicone rubber tube, leaving it there for a bit so it can soak up the toxins and then it is drained out; this is done four times a day, in between which the patient can go about their business as normal.

Perhaps even better than doing it during the day, there is a system where the patient connects themselves up to a machine overnight and, while asleep, the machine cycles the fluid in and out of the tummy. In the morning, it is disconnected and the day carries on as normal. This is the best form of dialysis for someone who is young and independent because it really means that they don't have to be tied down to hospitals or to a schedule during the day. Haemodialysis involves being connected up to a dialysis machine three times per week in a dialysis centre, and it usually takes about four hours, although it can be done at home. Under some circumstances, the patient can have a less complicated, more compact machine, so that they may connect themselves up and do the dialysis three times a week at home.

Dr Foggensteiner moved on to discuss transplantation and showed the delegates some slides of transplant operations, warning the squeamish to stare at their feet for a few minutes! He explained that kidneys come either from living donors such as parent to child, or sibling to sibling, or from those who have unfortunately died from a stroke or head injury, relying on the deceased's family agreeing to the donation. The advantage of a kidney transplant is that it frees the patient from dialysis completely, there is improved fitness, improved wellbeing and, in general, patients live longer with a transplant. It is also cheaper to transplant a patient than to dialyse and is therefore less of a burden on the NHS. It costs around £30,000 a year for dialysis, whereas a transplant operation costs £15,000, then £5,000 a year thereafter for drugs, so, not only is transplantation good for the patient, it is also good for the tax payer. However, there are problems and the main problems relate to drugs. The drugs have to be taken every day and they have to be taken reliably; if not, the body's immune system will reject the kidney. In addition, the drugs have side-effects and there is a small risk from the operation. The first transplant recorded was in France in 1906, and, rather ambitiously, the surgeons used a pig kidney but, of course, pigs and humans are somewhat different genetically and the pig kidney was rejected straight away. Scientists are still working on trying to perfect the use of pig kidneys but, at the moment, it is not an option. The first kidney transplant that really showed signs of promise was done some 50 years later, again in France, and the kidney was transplanted into the pelvis, a technique still in use today, so the technology of transplantation has not changed a great deal. What has changed enormously, though, is the drugs that are used, which are now much more sophisticated.

Dr Foggensteiner reported that, at present, they are doing just 1500 transplants a year; however, there are up to six thousand people waiting for a transplant, so you don't need to be a statistician to work out that there is going to be a bit of a wait. About 70% of transplants will last 10 years, so the outcomes are quite good. To sum up, he said, "Abnormal kidneys are common in BBS and it's important to monitor the kidney function and treat blood pressure, but, and this is an important 'but', very few patients do actually end up with kidney failure. It is important, however, that we diagnose and plan treatment early for everyone who's got Bardet-Biedl Syndrome, if only to confirm that the kidneys are working okay."

The Bardet-Biedl Brain Scans Project: first results and next questions

Dr Kate Baker
Academic Clinical Fellow
Developmental Cognitive Neuroscience Unit, UCL, Institute of Child Health

Kate is a junior paediatric doctor, who has previously carried out research about learning and emotional behavioural problems in other genetic syndromes. She is currently working with Professor Beales on a new project about Bardet-Biedl Syndrome and the brain. The aims of the project are to understand why some but not all patients have difficulties with learning, communication and emotional well-being. The project uses magnetic resonance imaging (MRI) to study the different aspects of brain development – something that has never been done before in BBS.

Those who were at last year's LMBBS Family Conference will remember the great talk Dr Baker gave about the brain and how it develops and about how she wanted to find the answers to the questions surrounding learning difficulties in LMBBS. It was great to welcome Dr Baker back to find out what she has been up to over the past year.

"I was here a year ago exactly and for me it has been an amazing year, with ups and downs but definitely a lot more ups than downs. I hope that you too have had more ups than downs this year. For those of you who didn't meet me or listen to me talk last year, there was one thing that I said at the very beginning of my talk last year, and it's probably the only really true thing that I said, and that was that you have the answers to the questions."

Over the past year, Dr Baker has conducted a study of the brain in those with LMBBS, looking for any differences that may account for some of the difficulties that those with the syndrome encounter. She had some fascinating results to share with everyone.

Dr Baker talked the delegates through various tests that had been done, all of which showed no difference between an LMBBS brain and a non-LMBBS brain. She showed the delegates scans, comparing a functional magnetic resonance imaging scan (fMRI) of the brain of someone with LMBBS with that of someone who doesn't have LMBBS, taken while they were thinking about words, and explained that again there was no difference between the two. Those with the syndrome use their brains for thinking about language in much the same way as those who don't have it. Dr Baker said there was lots and lots of thinking going in the LMBBS brains and the scans showed really great patterns of activation from the neuroscientist's point of view, maybe a bit more activation, using both sides of the brain. She explained that some might be using a bit more brain matter to do this kind of thinking than somebody who doesn't have the syndrome.

“So, we’ve looked at the overall structure of the LMBBS brain with our X-ray doctors and we can’t spot the difference. We’ve done some fancy functional MRI scans (and I’m quite pleased about this because I never thought there was going to be a difference on this one); it seems that, in terms of language and using both sides of the brain, overall there is not much difference. So was this a complete waste of time? Well, no, it really wasn’t because you were right.”

Dr Baker reported that they put together the scans of 10 LMBBS brains to create an average LMBBS brain and compared it with an average non-LMBBS brain and looked for shape differences and volume differences in the brain. At first, they didn’t find an awful lot. Dr Baker explained how she methodically analysed the data; however, it kept leading her back to the temporal lobe. She explained that she wasn’t expecting to find anything in the temporal lobe and thought any differences would be in the cerebellum and started to think that there was something wrong with the data.

Dr Baker said that she thought it was all rather strange until she remembered what the delegates had told her last year in the Conference Workshop about problems with memory. She then remembered some papers she had seen about mice with LMBBS and the cilia being necessary for maintaining the health of cells in an area of the temporal lobe called the hippocampus. Dr Baker explained that ‘Hippocampus’ means sea horse which refers to its curled up beautiful structure. It is connected to the anterior temporal lobe and is essential for forming new memories and accessing old memories that are stored in other parts of the brain.

Dr Baker continued, “So it came to me just like that, it must be something to do with hippocampus and I raced back to the lab and spent weeks and weeks measuring the hippocampus, tracing very carefully this beautiful structure in each of the scans of people with BBS and their matched participant controls. I thought ‘Gosh, if this doesn’t work, this is going to be weeks of my life.’ But this is what we do as scientists; we get a little hunch, and so far this is the strongest result I’ve had in my short career as a neuroscientist. So we don’t know whether the hippocampus works normally or not, but it’s much smaller in the participants who have taken part so far, which fits very well with the data for mice and it lends us some interesting hypotheses, interesting questions about what would link the genetics of BBS, the structure of the cilia, the maintenance of cells that turn over in the brain, and this particular part of the brain called the hippocampus, and how one is able to use that structure in the brain for forming new memories and thoughts through life.”

“So that’s where we have got to in this last year. And, for me, it’s been an amazing year, but the most amazing thing is I can come back and say to you, you were right, that I think you’re absolutely right about memory and that I think it has an impact on mood as well because you need to use your hippocampus to regulate your mood. And I hope to take this forward with some more scientists, but most of all take it forward with you so we can understand how the brain works differently in BBS, and also because I think, not just by understanding it, but by thinking about how we might be able to help you through life a little bit more, that this might be something that is of use to you in future.”

Kate finished her talk by inviting questions from the audience:

I’d just like to say a big thank you to Kate because Richard, our son, spent a very enjoyable day having his brain scanned and it was more like a visit to a theme park than a medical appointment, so he had a great day. But my question is about memory

because Richard has got just amazing retention and memory so I'm just wondering, would these results also explain excessively good memory?

"I'm really glad you asked the question. I was abbreviating really the whole of the experience at the workshop last year by saying there were problems with memory. So really I think one of the messages that came out was there seemed to be strengths in aspects of memory; for example, remembering phone numbers or remembering facts from a long time ago that other people have forgotten, but also a difficulty in acquiring new information and retaining things that had only happened very recently. Again, everybody in the group was a bit different but that was the sort of pattern that came out. So I think it's not just that overall there's a major problem with memory, but aspects of the way the memory system is working might be different."

Transition Options after School

Roger Simmons
Team Manager, Sheffield Futures

Roger has worked with young disabled people since 1977 when he became a volunteer at Sheffield Mencap's Gateway Club. In 1986, he qualified and embarked on a full-time career in youth work, working for the Sheffield Futures Stepping Stone Project, a city wide youth work project working to promote the inclusion of young disabled people. With the merger of the Youth Service and Connexions Service in Sheffield in 2002, Roger became manager of an integrated team of 16 staff, some youth workers, some Connexions advisers, all working on initiatives to support young disabled people in their personal development and in their transition from school to the adult world. Young people face a maze of options and difficulties at this stage in their lives and Roger's talk was about the options available and some of the changes that are taking place.

"When considering Sixth Form as the next step, it is important to think about the suitability of the school and whether it meets your particular needs. Can appropriate support be given in a sixth form or maybe a specialist sixth form in a special school? Is the curriculum appropriate? Are you going to end up with the qualifications or the experience you need for the next step in your life? What will be the outcome? Will it be A-levels? A-levels, at one time, were the be all and end all of education, but there are now many more routes that can be taken in education than the conventional A-level and degree course."

"When considering the local Further Education College, you need to look at what courses are available and whether specific needs can be met. If you have a visual impairment, will the support be there in a local college? Sometimes it can be, sometimes not. Once you have been through that course, what happens next, what do you do after that?"

"Some young people do go straight into work, but is that the right step? Is work what you want without any further qualifications or training? You work in the chip shop, delivering leaflets, things like that. It will get you a bit of money in, but is it going to set you on a life career that is going to last you and allow you to carry on learning and improving? Again, in the workplace, can appropriate support be given? Support for people in the work place comes from various areas such as the Department of Work and Pensions, Job Centre Plus and Job Steps. Another option is work-based training, which means getting into a work situation but with some training involved, which is going to improve your opportunities. There are mainstream and targeted options for work-based training. The questions to ask are: is this choice appropriate to my ability? Am I going to succeed? You also need to look for the appropriate support in either mainstream or a targeted placement. How long does the

scheme last? For example, “Entry to Employment“ schemes generally last for 22 weeks and formally measured progress is expected within this time. Some people with a significant learning disability will find it difficult to show that kind of progress within this time frame.”

“Moving on to residential specialist college, advantages and disadvantages: the whole life experience of going away from home, out of your comfort zone, is much the same as anybody would get when going to university; you get out of the family home and learn to live by yourself. This is what we term the 24 hour curriculum. In addition to the academic timetable, you will learn to make your own meals and things that are practical and useful in life. There is also the social life; anybody who has been to residential college will agree that it is an important part and sometimes missed out on if you have a day placement. Residential College also provides family respite from the general day-to-day caring, which is not to do with the individual but it is a by-product of the situation. Things to consider with residential specialist college is whether the mobility/independence training and social benefits, as well as the educational aspect have prepared you for when you leave – are all the skills transferable or will you have to start again? With day-placements at a specialist college, there is all the daily travel to deal with, however home life continues and you maintain local contacts. You’ll also learn your mobility locally.”

Roger moved on to discuss funding and how there are often ‘hoops’ to jump through to get funding for a specialist college placement. From a social healthcare perspective, funding methods are changing and there is a move towards giving people as much independence and control over their own lives as possible.

“The move is away from institutions and residences, and training centres generally and looking instead at individuals and supporting them with their particular needs, with a more personalised funding, tailor-made to the individuals concerned. This is called ‘Self-directed Support’, saying “What do you want? How much money do you think you need? We’ll decide we’re going to give you this amount. You decide how you’re going to use it”. It’s not quite as simple as that but that’s the general theme, a pooling of budgets. Pilot schemes have been successful in Sheffield and one that I was particularly interested in was where they paid a brother to buy a season ticket for Sheffield Wednesday, so he could take his disabled brother every week to watch football. So there you’ve got an afternoon of care for 23 weeks a year, which was an inventive use of spending money, and very cost effective.”

As mentioned previously, changes have taken place regarding funding and how you apply for it. Roger outlined the previous system and talked everyone through the new funding process.

“If I had been here last year, I would have said ‘You make an application to the Learning and Skills Council who hold the money up and down the country.’ Now that has disappeared. As of 31st March 2010, the responsibility for funding specialist provision lies with local authorities. Now we’re told the Young People’s Learning Agency (YPLA), will have the same amount of money as the Learning and Skills Council had and things shouldn’t change, however it is a local authority with squeezed budgets and squeezed resources so we’re waiting to see on that one. If you are looking at the specialist college route, it needs to be discussed pretty early, from year nine onwards at age 14. The family should visit the college, to see whether they like it and whether it can meet their needs. If so, the college will do a formal assessment to determine whether they can offer the service and if that is the case, they will offer a placement. You then make the application for funding or go to your Connexions worker.”

“The main criterion for this decision would be, ‘can your needs be met locally?’ If the local college says ‘Yes, we can meet that person’s needs’, the prospect of getting funding from the local authority/ YPLA is very difficult because they will say ‘We’re not going to spend the

£50,000 or £60,000 a year to put you in a specialist college if the local college, with some support, can meet your needs'. This can become a bit of a battleground because families, quite rightly, want what they feel is best, and the Connexions/Careers Advisor can provide support with that. They are not in charge of the funding but they can support you to put the application in. So, as I said before, it's a local authority decision."

Roger brought his talk to an end by discussing the role of the Connexions advisors, or Careers Advisors as they are sometimes called.

"They will be present at the annual review at school onwards, from year nine and they are your advocate. They will also help with careers interviews, support your funding applications and they'll liaise with the benefit agencies. They have a wealth of information about local options within the community, work, training and that sort of thing and are a valuable resource for you to call on."

Roger finished his talk by passing on some useful web addresses:

Young People's Learning Agency: www.ypla.gov.uk
NATSPEC (Specialist Colleges Association): www.natspec.org.uk
Connexions: www.connexions-direct.com
Department for Work and Pensions: www.dwp.gov.uk

An Update of the Lung Function Study

Dr Mathew Hind
Consultant Respiratory Physician, Royal Brompton Hospital

Dr Mathew Hind qualified at King's College London in 1995. He trained in general and respiratory medicine at the Royal Free, Hammersmith, St.Mary's, St George's and Royal Brompton Hospitals. He was a Wellcome Trust Clinical Training Fellow looking at the role of retinoids in lung development and regeneration in Professor Malcolm Maden's laboratory at the MRC Centre for Developmental Neurobiology at Guys' Campus, King's College, London.

Dr Hind has subsequent Wellcome Trust funding to investigate lung structure maintenance programmes. He is lead for stem and regenerative therapies for the advanced lung diseases unit, a partnership between Royal Brompton and Imperial College, funded by the National Institute for Health Research (NIHR). He has also established a lung regeneration laboratory at the National Heart and Lung Institute (NHLI) to investigate the cellular and molecular mechanisms of lung patterning, development and regeneration. Dr Hind offers a tertiary service for patients with lung failure, sleep and ventilation disorders and advanced chronic obstructive pulmonary disease (COPD). Dr Hind has been recognised for his work with awards from the British Thoracic Society and the European Respiratory Society.

"My name is Mathew and I'm one of the chest physicians at the Royal Brompton Hospital. I'm a scientist who's very interested in lung development and regeneration; however I also have a big clinical practice in sleep and breathing, which I think might be quite important to many of you with LMBB Syndrome. I think this is something that we haven't really touched on in the BBS field before and so what I'd like to do is present some of the data generated at last year's Conference which made its way to me as a clinician".

“Respiratory disease in BBS, why do we think that there’s an issue here? Why do we think it’s worth having a look? It’s because you told us last year that you have very frequent coughs, colds, asthma and wheeze, particularly in the winter, so we think this is something we should be interested in. We looked at some BBS mice and they appeared to have some lung problems, so we are very keen to find out if patients with BBS also have lung problems and we are setting up a study at the moment.”

“So what are the lungs? What do we know about breathing? Well, we all do it, don’t we, without even thinking about it. It’s very, very important. If we don’t breathe, we don’t survive. First, the body breathes in the air which is sucked through the nose or mouth and travels back through a series of tubes to the posterior part of our pharynx, which is the floppy bit at the base of the tongue. The larynx, or ‘Adam’s Apple’, is the start of the armour-plated trachea; this is a protective tube that can’t collapse. The trachea goes down into the middle of your chest and divides into two tubes called bronchi. These carry air into each lung. Inside the lung, the tubes divide into smaller and smaller tubes and at the end of each of these tubes are small air balloons called alveoli and it is here that the process of gas exchange takes place. This is a passive process, involving oxygen moving across the cells and carbon dioxide being released from them. This process is dependent, crucially dependent, on the surface area of the lung. Instead of having a huge, big chest, we have a small chest, with a kind of scrunched-up lung that has the surface area of a tennis court, which is pretty amazing.”

“Respiratory disease is common; we’ve all had coughs and colds, breathlessness, sputum production, wheeze, but also snoring, sleepiness during the day, and an inability to get going first thing in the morning, maybe with a headache, a bit like a hangover really. They’re very common symptoms of respiratory disease and I think BBS patients often have these symptoms. So what did we find in the mouse? Well, it looks like some of the BBS mice don’t seem to develop their lungs as normal mice do, and it seems the gas-exchanging surface area of the lung is reduced, so they have fewer but bigger alveoli. So what did we find at the conference last year? Many of you came in to have a chat with Robin (Quinlan) and she did some blowing tests. We found that many of the patients with BBS have a reduced lung function. There are two measures of lung function, the vital capacity, the amount of air that you can breathe out until your lungs are empty, and the FEV1, the amount of air that you can breathe out in a short period of time and both of those parameters were reduced in patients with BBS. Also, from talking to you, we’ve found that symptoms of obstructive sleep apnoea seem to be very, very common, and I’ll come onto that a little bit later. Interestingly, there was enough data to offer some screening tests at the Royal Brompton Hospital, to really investigate some of these symptoms in some more detail.”

Doctor Hind showed the delegates the data collected at last year’s conference which showed reduced values for the FEV1 and FVC tests. He was keen to state that he doesn’t think there is anything serious going on with the lungs in those with LMBBS, however there is ‘something’ going on. Dr Hind explained the process of diagnosing respiratory disease. A history is taken, noting frequency of symptoms such as coughs, colds, daytime sleepiness and whether there have been regular doses of antibiotics. Breathing tests, CT scans and echocardiograms are also used to determine lung function. They also do something called a sleep study, which investigates problems such as snoring, stopping breathing at night, waking up with a headache and generally feeling rotten first thing in the morning. Dr Hind explained further:

“Obstructive sleep apnoea is something that’s very important and very common in the general population. Up to four per cent of men and two per cent of women over the age of 50 have symptoms of obstructive sleep apnoea. Sleep apnoea is when the floppy bit at the top of our breathing tubes, just below our nose and our mouth, collapses back and stops us breathing during the night. The consequence of this is a loss of gas exchange and so the

oxygen levels in the blood go down. Fortunately, the brain wakes you up and says ‘Hey, hey, there’s something wrong’ and in the morning you wake up and think, ‘Did I go to sleep last night? My sleep was terrible. I don’t feel refreshed at all’; you feel like someone has been waking you up very, very frequently throughout the night. We think this is very, very common, and I think it’s probably more common in people with LMBBS. Although there seem to be some other factors involved, we think hormones are important; however we know one of the major factors of developing obstructive sleep apnoea relates to weight gain.”

“So what are the symptoms? I’ve mentioned snoring, unrefreshing sleep; you wake up and feel like you haven’t slept at all, daytime sleepiness and nocturnal choking. Your partner may say ‘Well, actually I was quite worried because you stopped breathing last night.’ Sometimes you get up to go to the loo many, many times, maybe three or four times at night, and that’s not normal but it’s a very common symptom of this condition. Morning headaches as well, feeling like you’ve been out drinking when you haven’t had anything to drink at all and reduced libido. How do we measure sleepiness? Well, we’ve devised this tool called the Epworth Sleepiness Scale. There are seven questions and all you have to do is indicate the chances of falling asleep in certain situations: no chance, slight chance, moderate chance or high chance. The situations are: sitting reading, watching TV, sitting in a public place like a theatre, or a cinema, or something like that, a passenger in a car for an hour or lying down to rest in the afternoon. If you’ve ever fallen asleep when you’re sat talking to someone, that really suggests that you’ve got this condition. What about sitting quietly after lunch with no alcohol? If you answer yes to these, you could have a problem.”

“So what do we do in a sleep study? Well, we measure your oxygen level when you’re asleep. We also have a microphone to measure your snoring and we look at your body position. When you’re snoring and your airway is obstructing, your heart rate goes up and also your blood pressure goes up. It’s a very, very common cause of what doctors call difficult blood pressure, difficult hypertension, where you might need one or two different types of treatment for blood pressure. So what are the complications of this very common syndrome, obstructive sleep apnoea? Well, they’re multitude really, sleepiness, impaired concentration and poor memory. We heard from Kate earlier about changes in the brain. We also see these changes in the brain in the hippocampus in people who have sleep apnoea, severe sleep apnoea. So I wonder if there’s a link there. Sleep apnoea can affect the heart and the lungs. It can also affect diabetes. It gives you enuresis, so you get up in the night to go to the loo, and it can also cause changes in your blood count as well, so there are lots and lots of medical things that doctors are worried about with regard to obstructive sleep apnoea.”

“The reason I have highlighted sleep apnoea in particular is because there is a very straightforward treatment called CPAP (Continuous Positive Airway Pressure). It’s a little machine that you wear that blows air in through your nose or through your mouth and nose and holds the floppy upper pharynx at the back of your throat open so that you’re not trying to work hard against a closed upper airway. You don’t have to have a big scary full face mask; you can have a nasal mask or mouth mask or even little nasal pillows that rest inside your nostrils that blow in air to hold the pharynx open. It restores normal sleep by preventing your oxygen levels from falling which, in turn, prevents you from waking up at night. It is a very good treatment. I’m a big fan of it because you abolish snoring, your sleep becomes refreshing, you’re no longer falling asleep during the day, this frightening nocturnal choking disappears, your headaches can resolve and your libido can improve.”

Professor Beales thanked Doctor Hind and invited questions:

Is it difficult to get a CPAP machine, are they expensive?

"They don't cost you anything if you need one. If you're sleepy during the day, and you snore and you have symptoms suggestive of sleep apnoea, go and see your GP for a referral. You need to be seen in a sleep clinic, which are dotted around the country. You will have to have a sleep study; however we now run studies in the patient's own home. You will be shown how to use a diagnostic kit, which you then take home with you. When you come back, we download the information and we will then be in a position to say 'Yes, you have sleep apnoea,' or 'No, we need to find a different cause of your sleepiness.' There are other causes of sleepiness that we need to think about, but by far the most common is sleep apnoea, and if you have sleep apnoea, based on the results of the results of that sleep study, you'll be offered a CPAP."

Blind Football Academy
Jo Powell – Royal National College for the Blind (RNC)
Ajmal Ahmed – England Player

The Royal National College for the Blind (RNC) is the UK's leading specialist residential college offering further education and training for people with sight loss from age 16+. Along with a wide variety of academic and vocational programmes, the College now runs the world's first Blind Football Academy, where students can train to play blind or partially sighted football at local, regional and national levels. RNC has some of the best facilities in the country for disability sports and has been selected as the host venue for the 2010 World Blind Football Championship.

Jo Powell: "Hello, my name is Jo Powell and I'm from the Royal National College for the Blind, which is one of the specialist residential colleges Roger spoke about earlier. We offer education and training for people who come from all over the country and abroad to us in Hereford from the age of 16 upwards. In previous years, our Head of Marketing, Cathy Fletcher, has given a talk about the RNC; however, we thought for this year, because this is a special year for the college, we'd give you something a little bit different. I don't know how many of you know about the sport of blind football, but this summer the World Blind Football Championships is going to be held at the college in Hereford, so this is a big deal for the college and, we hope you'll agree, it's a big deal for the VI community in the UK as a whole. We're going to be hosting the largest disability football event that's ever been held in the UK, and we thought we'd come along today, myself and my colleague Ajmal Ahmed, who has actually been a captain of the England blind football team, and tell you a little bit about blind football."

England vs Spain - Photo courtesy of the FA

"Blind Football is a very fast five-a-side game, involving serious ball control skills. There are four blind players who have to wear blindfolds, not only to protect their eyes but also to ensure that everyone is playing on the same field. No-one can see, no-one can cheat. Goalies are sighted and they're allowed to shout so everyone can hear from a directional point of view, and the ball is heavy and has ball bearings inside it so you can hear it rattling when it moves. A couple of years ago, 'Inside

Sport', the BBC's sports programme, came and filmed at the college. The reporter, Des Kelly, had a go at blind football, which I think is brilliant, it's the best way to find out how incredible the sport is. Just put a blindfold on and have a go yourself, running up and down a blind football pitch, and then you'll start to understand what an incredible sport it is."

Jo showed the delegates the 'Inside Sport' footage which is awe-inspiring. To view the film online, type 'Inside Sport – Blind football' into the search engine and it should come up.

"The film gives a little bit of a flavour of blind football and this film was taken just before the guys went off to Beijing in the summer of 2008. You can probably agree that David Beckham would give anything to have captained the England team to be ranked fifth in the world, but this is exactly what Ajmal has just done. I will hand over to Aj now and he can tell you a bit about his experiences of playing for the team, how he got into playing blind football, and what it was like to play in Beijing against the world's blind footballers."

Ajmal Ahmed: "I've been playing football now for years and I'll tell you briefly how I got involved with football. I come from Liverpool, where everyone plays football. I think growing up on a council estate, you do two things in Liverpool as a kid, you play football or rob cars and I wasn't very good at the second one. So, yes, from a very early age, my only ambition was to play football, but even then I thought, well, at the time I thought, it was just a pipe dream because blind football didn't exist."

"I went to RNC in '98 to do my A-levels and I remember one of the first people I met for my induction was Tony, who's the manager, and he was telling us all about the recreational stuff that the college do and said 'Oh, we've got a blind football team and an international team as well', and from that moment I thought to myself, 'I'm definitely going to get on that team'. Within four months, I went on trials and, within nine months, I was swanning off to my first European Championships. Since then I've played in 10 major events - three world cups, six European Championships and, of course, I was lucky enough to captain the team to Beijing."

"People always ask 'what was the experience like', but, when I went there, I didn't really take it in because I didn't want to be overawed by the fact that I was playing in the first Paralympics that the England blind team had played in, so I sort of went on autopilot; you have to be professional about it. But I've finished playing now, and I look back at it and just think about the organisation, of being out with the team for a month with all the sports scientists and the physio's. The Olympic Village was full of thousands of world class athletes at the top of their game who were just unbelievable, and even thinking about walking out with the team with the armband on my arm, with 5000 people cheering or booing, it just sends a shiver up my spine. I think the only thing that I'm sorry for is that I've retired now. It's a bit of a shame because blind football is growing every single year, and even more so now we've got the World Cup in our own country this year, and then 2012, and I'd have given anything to be a part of that. But it is growing and I think we've got a really good chance this year. We've got a mix of young lads, some experienced old time campaigners, so, especially being on home soil, if we have the support, there's no reason why we can't go really far."

Jo finished the talk by inviting everyone to go along to Hereford in August to watch the World Cup Blind Football Championships. We wish the England team, Ajmal and everyone else at the RNC every success in the future.

Annual General Meeting
The Hilton Hotel, Northampton
Saturday 24th April 2010

Minutes of previous Annual General Meeting
The Minutes of the Annual General Meeting on Saturday 18th April 2009, previously circulated, were agreed and signed.

Election of Officers
The current officers, Phil Humphreys (Chairman), Terry Crotty (Vice Chairman), Julie Sales (Secretary), Kevin Sales (Treasurer), Chris Humphreys (Conference and National Co-ordinator), Anne Crotty (Fundraising Co-ordinator) and Tonia Hymers (Newsletter Editor) were all eligible for re-election and were duly elected unopposed.

Election of Committee
The current committee members, Steve Burge and Allan Clark have a further year to serve on the committee. Tina Hickey and Julian Thomas, having served two years retire this year but are eligible for re-election. Nominations were received for Tina Hickey and Richard Zimbler. In the absence of any further nominations, the Society are delighted that Tina was re-elected and Richard was elected to the committee unopposed.

Chairman's Report 2010
It has been a busy and exciting year for LMBBS and our congratulations go to Professor Beales and teams for their successful application for funding to set up dedicated LMBBS Clinics. The first clinic was at Great Ormond Street Hospital on 16th April, 2010. You should have all received information regarding the clinics by letter and further information will be in the newsletter, which is being printed as we speak. On behalf of the committee and myself, I would like to thank Tonia, Julie and Chris, who will take on the roles of Clinic Co-ordination and Family Support, for their immense input over the past several months; there has been an incredible amount of preparation to have everything in place for 1st April and this is in addition to their everyday committee roles. Thank you also to our very stressed Treasurer for his role in dealing with the Finances for the clinics budget.

You will know, from letters received, that we have updated our data base in line with the Data Protection Act. All our members have been contacted and asked if they wished to remain on the database and to update any information. If you change your address, phone number or e-mail address, please inform Julie, Tonia, or Chris, so that we can adjust our records.

LMBBS is one of the founder members of the newly formed 'Ciliopathies Alliance UK', along with Professor Beales, Dr Dan Jagger, Dr Claire Hogg, Hannah Mitchison, Polycystic Kidney Disease and Primary Ciliary Dyskinaesia support groups and Alstrom UK. The launch date is set for 29th November this year.

Tina Hickey is currently working with Professor Tim Barrett of Birmingham Children's Hospital, and Kay Parkinson of Alstrom UK on the dissemination of characteristics of LMBBS and Alstrom Syndrome for EURORDIS. This is the voice of rare disease patients in Europe, an alliance of patients associations dedicated to improving the quality of life of all people

living with rare diseases in Europe. There will be no financial cost to the society. We will bring you more news on this as we progress.

The Genetic Interest Group Rare Disease Day was well represented this year for LMBBS, by Allan Clark, Pauline and Peter Taylor, with their son Keiran, for Scotland, Jonny and Sharon Fegan for Northern Ireland and Chris for Wales; (there was no meeting in London this year because of the impending elections). Chris was asked to speak at the Welsh Assembly in Cardiff on 'Patients & Researchers: Partners for Life', about the need for research into rare conditions and development of treatment and support for people living with them. This provided an opportunity to speak of the partnerships we have forged with many eminent clinicians over the years and the introduction of LMBBS Clinics in 2010. As a result of Chris's consultations with the Commissioner of Health for Wales, I am pleased to say the Welsh Assembly have not only agreed to fund LMBBS Clinics for Welsh patients, (funding is now available on request), but all other Rare Diseases will have the same opportunity to access Clinics of Excellence outside Wales.

Thank you to Julie and Lance for the newly designed web page. There will be an opportunity for members to contact each other and visit the LMBBS shop to purchase our merchandise. At the end of 2009, we had 11,848 unique visits, 19,407 return visits and a total of 160,255 hits. The USA is still the most popular visitor, followed by the UK. We now have 300 families and 178 professionals on our database, which is being updated. Thank you also for your work as Conference Childcare Co-ordinator, Julie.

Thank you to our Fund Raising Co-ordinator, Anne Crotty, for her continuous efforts to gain funding, although it has been a very difficult year, with a positive response from only two out of nine applications submitted. (Grant-making bodies have been affected by the recession as well and have limited funds to share out.) We thank our members for their fund raising and the Yapp Charitable Trust for their generous grant of £3,000. In addition, we have been allocated a Radio 4 Appeal on 27th June, 2010. The Super Grand Sports Raffle and National Coffee Morning were successful enough to encourage us to plan the "Big S", a 'Summer Sundae' of fundraising events in 2010. Please speak to Anne if you think you can help with this event.

Thank you to our Newsletter Editor, Tonia Hymers, who has had to combine her final year of studies with family, clinics, newsletter and also her role of Crèche Co-ordinator for the conference. Remember that, without your photographs and snippets of information, there will be no newsletter. Tonia can be contacted by e-mail at toniahymers@btinternet.com.

Thanks to Chris for her work as National Co-ordinator, often the first point of contact for our members and professionals, and for her continued role of Conference Co-ordinator.

Thank you, once again, to Jonny and Sharon (Fegan) and family and friends, and the people of Newry, for inviting James and myself for a wonderful weekend of fun and fund raising at the end of August 2009. An incredible £3,482 was raised throughout the weekend.

Thank you to all those who have raised money for the Society this year; your donations have been very much appreciated, for without you the Society will not survive

Thank you to Allan Clark for representing the Society in Sight Village, Scotland. James and I attended Sight Village, London. If there is anyone here who would like to help at Sight Village, Birmingham in July this year, please see one of the committee for details. Although it is a three day event, if you can spare a day or even part of a day, your support would be very much appreciated.

Thank you to Ryan Jones, our Patron, for his continued support for the Society. Ryan recently donated a 'signed training squad jersey' with two signed pairs of shorts that he wore for the Six Nations Cup; they were auctioned locally at a charity golf tournament for the Society and raised £1,040. He is unable to be with us today as he is playing in Ireland.

To Professor Beales, Honorary Officers and members of the committee, thank you for your support and sterling work over the past year.

Our aims and objectives for the coming year are as follows:

- To continue to actively seek sponsors to fund future conferences.
- To continue to support Professor Beales & Teams in the Designated LMBBS Clinics in Birmingham and London.
- To finalise the update of the LMBBS Database, checking that the information we hold is accurate, in line with the Data Protection Act.
- To move forward in our involvement with the Ciliopathies Alliance Group.
- To continue our involvement with EURORDIS.
- To build on the success of national fundraising with the Summer 'Big S', making it a significant annual Event, raising awareness and funds.
- To continue fundraising, and to support our members in their fundraising.
- To continue to produce Newsletters and an annual Conference Report.

We will strive to achieve these aims and will, as always, work together to ensure that the Society continues to go from strength to strength.

Before I end, I feel I must mention a very special young man, Shane Ryan, from Southern Ireland, who has been winning trophies in competitions for Adaptive Rowers. It really is a great sport for anyone with a visual impairment - most rowing clubs now include adaptive teams as it is now a sport in the Paralympics. There is lots of training and discipline involved which suits him down to the ground. It has also done wonders for his weight and blood pressure - his doctor is delighted with him. The latest news from the Ryan family is that Shane won 1st place at the Indoor Rowing in Boston. Their next trip is to Italy to an Adaptive Regatta in Varese. I think you will agree that he is an inspiration to all.

Treasurer's Report

The Society's accounts have been balanced and audited by our accountants, Michael Thompson and Co in Maidstone, for the 2009 tax year. Unfortunately, compared to our record year in 2008, 2009 has not been that good and we made a hefty financial loss of over sixteen thousand pounds. Obviously, with the current economic climate, that is probably no surprise. Things are not looking that rosy this year, either, and we therefore have a very tough year ahead, so any fundraising that we can do as a charity within the next year will be very much appreciated. Otherwise, the Family Conference next year will have to be a day event, rather than the weekend event that everybody enjoys.

Appointment of Auditor

The Chairman confirmed that Michael Thompson has kindly agreed to continue as Auditor for another year and was duly re-appointed.

Any Other Business

Amendments to the constitution – Office of the Scottish Charity Regulator (OSCR).
In order to help our Scottish members and friends to carry out fund raising activities in Scotland, it is necessary for our Society to register with the Office of the Scottish Charity Regulator. We cannot do this with our constitution as it stands. However, all that is required by the OSCR is that we amend it slightly by adding a paragraph as outlined below. Clause 14 should now read:

"DISSOLUTION

(a)If the Committee by simple majority decide at any time that on the ground of expense or otherwise it is necessary or advisable to dissolve the Society, it shall call a meeting of all members of the Society who have the power to vote, of which meeting not less than 21 days' notice (stating the terms of the Resolution to be proposed thereat) shall be given. If such decision shall be confirmed by a simple (two thirds) majority of those present and voting at such meeting the Committee shall have the power to dispose of any assets held by or on behalf of the Society. Any assets remaining after the satisfaction of any proper debts and liabilities shall be given or transferred to such other charitable institution or institutions having objects similar to the objects of the Society as the Committee may determine but if and so far as effect cannot be given to this provision then to some other charitable institution or institutions.

(b)Nothing in this Constitution shall authorise an application of the property of the charity for purposes which are not charitable in accordance with section 7 Charities and Trustee Investment (Scotland) Act 2005."

As there were no objections, the amendment was accepted by the meeting.

In the absence of any further business or questions, the meeting was closed.

Your Committee for 2010-2011

Phil Humphreys - Chairman
Phil is married to Chris, our Conference & National Co-ordinator, and together they have five children and three grandchildren. As foster carers with 42 years' experience, they have adopted three children and are currently fostering a 15 month old girl and 3 year old boy. Phil worked in South Wales & Gwent Police Force for thirty years, before retiring in 1992. Since then, Phil has worked in residential homes for adults with learning difficulties and has been a primary school caretaker. More recently, he was a driver/escort for schoolchildren with additional needs, before retiring in 2009. Phil has been a member of the Gwent Police Choir since 1971 and is currently the Committee Chairman. His involvement with the LMBB Society came in 1995, with the diagnosis of his son. Phil joined the LMBBS Committee in 2002 and became Chairman in 2003. Phil is a Director of LMBBS Clinics Ltd (non-remunerative) and is also a member of the LMBBS Finance Sub-Committee.

Terry Crotty - Vice-Chairman
Terry Crotty has been Vice-Chairman of the Society since 2006. He is a member of the Finance Sub-committee of the LMBBS Management Committee. He is a retired Fire Officer and, together with Anne, his wife, became involved with the Society soon after the diagnosis of their grandson in 1997. He was a member of the local Lions Club for several years and has been actively involved with his local church. Terry and Anne represented the Society at the Sight Village Exhibition in Birmingham for several years and have now become directors of LMBBS Clinics Ltd (non-remunerative).

Kevin Sales - Treasurer
Kevin is married to Julie, our Secretary, and together they have two daughters, both of whom have LMBBS. They attended their first Conference in 1998, following their children's diagnosis. Kevin worked in the Pensions Industry for 24 years until being made redundant in 2003. This allowed him to retrain as a bookkeeper which is his current employment. Kevin also works as a Student Support worker at a local College for the Visually Impaired. Kevin joined the LMBBS Committee in 1999 and became Treasurer in 2000. He is a Director of LMBBS Clinics Ltd (Non-remunerative) and is also the Company's Book keeper. Kevin also Chairs the Finance sub-committee of the LMBBS management committee.

Chris Humphreys - National and Conference Co-ordinator
Chris is married to our Chairman, Phil, and together they have five children and three grandchildren. As foster carers with 42 years' experience, they have adopted three children and are currently fostering a 15 month old girl and 3 year old boy. Chris worked in the South Wales Police Force, where she met Phil. She became Officer in Charge of a Children's Home, before leaving to focus on fostering and childminding. With the arrival of James, a 3 week old baby, life changed, and Chris's involvement with LMBBS came in 1995 when, at 13 years old, James was finally diagnosed with the syndrome. Chris became a committee member in 1996 as Fundraising Officer and later as Secretary, before her current role as Conference & National Co-ordinator. Chris is also a Director of LMBBS Clinics Ltd (non-remunerative). More recently, Chris works with prospective new carers as a member of the Social Services Fostering Team. Chris has been working with GIG (Wales), Ciliopathy Alliance UK and is the LMBBS Clinic Family Support Worker at University Hospital and Children's Hospital, Birmingham.

Julie Sales - Secretary
Julie is married to Kevin, our Treasurer, and together they have two daughters with LMBBS. They were made aware of the charity when the girls were diagnosed in 1997 and first became involved on the committee as observers in 1998. Julie has worked in various professions, including secretarial, catering, physiotherapy, and learning support. She has undertaken independent study with the Open University and has completed a counselling course. Julie is now the LMBBS Family Support Worker, co-ordinating BBS clinics, at Guys Hospital, London and is a director of LMBBS Clinics Limited (Non-remunerative). Julie joined the LMBBS Committee in 1999 and has been Secretary for five years. She is Childcare Co-ordinator for the Conference Weekend Outing and has previously helped with the Society's Helpline.

Anne Crotty - Fundraising Co-ordinator
Anne Crotty has been Fundraising Co-ordinator for the Society since 2004. She is a retired Midwife Teacher and, together with Terry, her husband, became involved with the Society soon after the diagnosis of their grandson in 1997. She spent the first half of her life travelling from place to place as her father and her

first husband were in the Army. She has served as a Steward for the Royal College of Midwives and as an Elder of her local church. Anne and Terry have represented the Society at the Sight Village Exhibition in Birmingham for several years and have now become directors of LMBBS Clinics Ltd (non-remunerative).

Tonia Hymers - Newsletter Editor

Tonia is married to Rob and together they have two sons, aged 10 and 14, the oldest of whom has LMBBS. They went to their first Conference in 1998 and were both coerced onto the Committee soon after that. Tonia was Fundraising Co-ordinator for five years, before taking on the role of Newsletter Editor. She is also the Conference Childcare Co-ordinator for the Creche. Tonia worked for Barclays Bank for fifteen years before leaving to concentrate on raising her family. In 2005, she enrolled with the Open University and in, 2010, she completed her studies, gaining a BSc in Social Sciences. In April 2010, Tonia became the Admin/Clinic Co-ordinator for LMBBS Clinics Ltd and the Family Support Worker for BBS Clinics at Great Ormond Street Hospital, London. Tonia is also a Director of LMBBS Clinics Limited (non-remunerative).

Tina Hickey

Tina is married to David and they have two children, James who is 19 and Angharad aged 9. Geneticists expressed concerns regarding some of the characteristics James displayed fairly early on and they were given a tentative diagnosis of LMBBS. Tina and David made contact with the LMBBS family support group whose expertise and friendly ear, they say, helped them through the uncertainty of those early years. Tina has been involved with the Society since 1993, soon after James was diagnosed, and returned to serve as a committee member in 2006. Tina qualified as a Registered General Nurse in 1984 and has worked in various nursing roles, mainly in Oncology/ Haematology. Tina and David have also been foster parents for social services and provided short term respite care.

Steve Burge

Steve was diagnosed with LMBBS at 11 years old, lost his vision at the age of 21 and has been involved with the Society since 1997. Music has always played a big part in Steve's life and he credits his love of music, especially his drumming in helping him cope with having the syndrome. In 2000 Steve qualified as a sound engineer from the Royal National College in Hereford and has since been involved in numerous professional recording projects, both as a drummer and as a sound engineer. Since 2006, Steve has been the Disability and inclusion director for RBCR Ltd a company set up to help give opportunities to blind and partially sighted people who want to pursue a career in radio broadcasting. More recently, in early 2010 Steve qualified as a teacher and has started a new business, Sunstream Multimedia.

Allan Clark

Allan Clark has been a committee member since 2009. Allan works for Macmillan Cancer Support in Scotland and manages a volunteer service, supporting those affected by cancer. He has three children with his wife Jacqueline and both his sons have LMBBS. Allan has been involved in campaigning with GIG in Scotland, was part of the school speaker programme for Jeans for Genes and is currently training to walk the 96 miles of the West Highland Way to raise funds for LMBBS.

Richard Zimbler

Richard was diagnosed with LMBBS when he was six years old. He went to Welshpool High School and continued his studies at the Royal National College for the Blind (RNC) in Hereford. Richard has worked for Disability Arts, Shropshire (Dash), Masquerade Integrated Drama and DD Aware Ltd on a voluntary basis and has been a Student Ambassador Volunteer for the Open University, Powys. He is a member of 'Look', the National Federation for Parents with Children who are Blind or Visually Impaired and has attended Sight Village with the group. Richard is also the Chair for Oldford Communities First. Richard is a performing artist and likes to act, sing and play the drums, both African hand drums and full kit. He was elected to the LMBBS Committee in 2010 and is committed to helping to further the aims of the Society and to helping those with a visual impairment.

What's the point of crying?

A Personal Perspective by Emma Turnbull

"Hi, I'm Emma. Chris asked if it would be possible for me to give a talk on what it is like with three children affected with Laurence-Moon-Bardet-Biedl Syndrome. We have now come to the point where we say, 'What is the point of crying, as you only end up with a headache, and the problem is still there'.

I was four months pregnant with Nikita in 1999 when I went to have my first scan. All excited, I went along to have the scan done, only for the sonographer's face to go white. At this point, I was ushered into a side room. After a lengthy wait, a doctor came to see me and explained that I needed to go to the Foetal Medicine Unit. That was it; he left the room with no more explanation. So nosey me went to the door and was stopped dead in my tracks by the conversation that was taking place in the corridor by the doctor, the sonographer, and a midwife. "We will book a termination for Miss Palmer". At that point, the tears started, and, might I add, many more were to come with the pregnancy. I went home feeling, not like a new pregnant mother, but at a loss to know what to do and what was happening. Paul was gutted.

Two days later, I was in a strange hospital at a unit which I knew nothing about. In walks a newly trained doctor called Professor Robson. He was to be a rock, throughout all my pregnancies, to us both. I was told that I had to have an Amniocentesis Test and Chorionic Villus Sampling. Both involved big needles. Whilst doing these tests, Professor Robson finally explained what was wrong at this point. He explained that the kidneys were pink and echogenic. There were six fingers and six toes and the diagnosis was LMBBS. At first I couldn't pronounce it, let alone understand it. Next, another room and more tissues for the tears. In comes a swarm of specialists, some with the most appalling bedside manners I have ever come across. After he introduced himself as one of the renal team, one of them said, 'If it is to be one of the lucky ones, it will die hours after being born'. Talk about being hit by a sledge hammer. After this sunk in, the anger set in, along with attitude. I stood up and said, 'As long as she has a heart beat she stays, and I don't want to see another box of hankies', and I walked out. At the geneticist appointment, we were more or less told to go home and deal with it.

I went to work that night, upset on the medical unit at what the doctor had said. He then asked what was wrong. I went on to explain everything. The next thing I knew, his desk was cleared of all paperwork, and the search for information was on. He found out the

contact number for the Society through Contact a Family. I went home a little easier. When you're pregnant, you have all the stresses of a normal pregnancy, sickness and emotions but, to have the stress of knowing that there's a medical problem with your child, it is ten times worse. D-Day, 25th December 1999 and Nikita is born after four days in labour. Yes, there are extra toes and fingers, but so what? We were told at a scan with the renal team she wouldn't be able to wee. The midwife picked Nikita up, only to be drenched in urine; she had done her first wee, and we were so proud.

Next came referral, referral, referral. It was a never ending stream of daily appointments. Ophthalmology, renal, heart, plastic surgery, dieticians, paediatrics, geneticists, test after test. Nikita didn't reach her milestones like a normal child. She was, and will always be, a little slower than others, but she gets there in the end. Nikita had her extra toes and fingers removed at thirteen months. To this day, she hates the surgeon who did the op. We have to sneak her in the back door so that she doesn't see the surgeon until it's too late. And they say children forget. No, they don't.
Nikita started to lose her eyesight at about sixteen months. We noticed with bumping into things. She hated the dark. Now she has difficulty with kerbstones. If she drops something, she feels for it, rather than looks for it. But this does not stop her from enjoying what she's doing. She adjusts quite quickly to her surroundings. It's very rare that she goes out at night, as she cannot see in the dark. Her kidneys are stable at the moment, along with her heart murmur. Weight is an extreme issue with Nikita. From the age of two, she seems to have always been on a diet and it can be hard when all the other children are getting treats and you have to say 'no' to Nikita.

Her speech was extremely poor in the beginning, but, with help from a Speech Therapist and Portage (a home visiting educational service for pre-school children with additional needs), Nikita's speech progressively improved over the years. Now the problem is getting her to shut up, because you can't get a word in. School was a problem at one point, due to her being bullied, but we resolved that by changing her school. She now attends a special school for media and arts. She tries her best at everything she does and that is all we can ask.

Nikita's brother, John James, came next and it was clear from the beginning of the pregnancy that he was going to be very demanding. For five months of the pregnancy, I was in hospital; if it wasn't for the four stone I dropped because of the sickness, it was the problem with my kidneys due to the dehydration. The four-monthly scan was a classic. By this time, I was automatically sent to the renal unit. Lying on the table having a scan done and the sonographer's face goes white again. She mumbles to another member of staff and her face goes white also. At this point, I ask what's wrong. A blank expression covered their faces. So I answered the question for them. The kidneys are pink, echogenic, and there are six toes and he has LMBBS. Their expression said it all. At this point they went away for a specialist. In walks Professor Robson. After a lapse of seven years, as soon as he walked in, a smile shot across his face, and he said, 'Here we go again'. The only difference with the specialist unit was that, this time, they removed all boxes of hankies, as this annoyed me. What is the point of crying? You only end up with a big headache and the medical problem is still there. I also managed to skip the big needles this time.

D-Day, 29th December 2007 and John James is born, weighing in at eight pounds four ounces. Yes, he did get stuck and he was premature. Referral, referral, referral. John James's kidneys had been badly damaged due to reflux. He has, over all, fifty per cent

kidney function. He needs permanent antibiotics to stop infections. His first appointment with the renal team was with the doctor with no bedside manner. Needless to say, when he realised who we were, an apology was the first thing we received at this appointment. We had great satisfaction in telling him that Nikita is still here and doing okay. He may be a brilliant renal surgeon, but, by God, his attitude sucks as Nikita tells him frequently. John James's eyesight is quite poor and our ophthalmologist picked this up straight away. In an attempt to help, John James was prescribed glasses. I never knew frames could contort into so many different shapes. He is only two and his cooperation with glasses is non-existent. He has no speech at the moment, which causes him great frustration but, with the help of Portage and Speech Therapy, we are teaching John James the Picture Exchange Communication System (PECS). For those of you who don't know, this is a system of pictures, which help the child to ask for something by handing you the picture. He's also under the dietician for his weight, which, like Nikita's, is a constant battle. John James is very much daddy's little boy. He's very emotional. We have decided not to have his extra toes removed; that is a choice for him.

Then along came a little sister for Nikita and John James. Summer was a very unexpected big surprise. I had had all the tests done for the menopause and I was told that I couldn't have any more children as I was through the change. Wrong. What a shock we got. Four monthly scan, same pained expression on sonographer's face. I answered all the questions for them. Pink echogenic kidneys, polydactyly and, again, the look of relief in their faces said it all. I then told them to send me down the corridor and tell Professor Robson I was back again. Sickness was bad again; six months in hospital with dehydration. I did manage to get home twice for three hours but then the sickness started again. I was that poorly whilst carrying Summer that Professor Robson stayed by my bedside for two nights in a row as, over the years, we had all become friends. He stressed that I would not be able to carry full term. Six and a half months into the pregnancy, Summer suffered severe difficulties in the womb. Needless to say, an emergency C Section was needed and she was born on 17th July 2009. She went straight into Intensive Care and was there for ten days. All appointments were made for us before we left the hospital. We are now the only family in the North East with block bookings for appointments, and this has taken two years for us to achieve.

It is not the end of the world when it comes to being diagnosed with a medical condition. Everybody is their own unique person. It doesn't matter how fast or slow they are at achieving their goals in life, as long as they've enjoyed trying, they will get there at their own pace. So many people ask us how we cope with all three having the syndrome. You have to. You have the setbacks of the scans and you just have to pick yourself up and dust yourself off. At the end of the day we have three beautiful children. So what if they are complicated? They're ours. We just take each day as it comes. No two days are the same."

A Great Weekend!

My name is Greg and I am fourteen years of age. I am into music and I play the drums. My sister has BBS and so my family decided to go to the LMBBS Family Conference. We hadn't been before, but I was told about people that were going, including a boy around my age called James, whose sister, like mine, has BBS. I was nervous at first but we had a friendly chat and got along really well. We went to breakfast together the next morning. After breakfast, we met the staff that would be keeping an eye on us at the theme park. They were very kind and made sure we had fun. We got onto the coach and met the other people that were in our group. They too were kind and funny and I had fun with them at the theme park even though, at the end of the day, I was extremely wet from the water rides. We got back to

the hotel, enjoyed a lovely dinner and then played some family games which were set-up in one of the conference rooms.

The next day we had a lie-in and then a later breakfast. I asked for pancakes and syrup which the hotel staff kindly cooked for me as it was off the menu. It was so good that I asked for seconds! Later me and James went swimming in the hotel pool, we had great fun. We also started chatting about our sisters' disabilities.
Unfortunately, it was soon time to head home so we said our goodbyes and farewells and packed up and headed off. I still keep in touch with James and his family on Facebook and hope to see them in the summer.

By Greg Dowswell

Fun and Fellowship



One of the highlights of our Annual Weekend was the Saturday evening, when the lobby of the hotel was filled with fun and fellowship. The youngsters rejoined their parents, the carers were able to relax and even the Committee were able to unwind to a great extent.

Our resident quizmaster, Dennis Clarke, produced another set of challenging questions. There was fierce rivalry between the teams and a great deal of fun was had by all. Thank you, Dennis and Fiona, for providing super entertainment and prizes.

We are very grateful to those of you who supplied raffle and tombola prizes. The tombola was enjoyed during the day by youngsters and adults alike. The raffle followed the quiz and our thanks go to the **Hilton Hotel** for their star prize of a Weekend for Two and to the Petty family for their HUGE box of goodies. Thank you, too, Ray and Doreen Larkin, for your time and energy and persuasive powers.

Although the Annual Weekend is not specifically a fundraising event, the generosity of those present enabled our Treasurer to go home with a smile on his face! As usual, we were given the proceeds of several collection boxes, always gratefully received. The quiet commitment of those of you who support the Society in this way is impressive. We could not manage without you. Business was brisk for most of the weekend at the merchandise table, for which, our grateful thanks to all who support the Society in this way.

We were also pleased to receive a cheque from Richard Zimbler who sang for us last year and subsequently sold his CDs for Society funds. Thank you, Richard.

During the past year, we received a lovely surprise from the **Great Gizmo Company.** It was a box of interesting toys and games. Those who opened it said it felt like Christmas! Thanks to their generosity, we were able to give the youngsters an additional treat before they went home.

One person who was missing over the Weekend was Louise Butcher, (one of our carers), who was running in the London Marathon on behalf of the Society. To quote Louise, she "was gutted" about not being there with us. However, out of sight was definitely not out of mind as Allan Clark went round "persuading" everyone to sponsor Louise. He even tackled the Middlesex Cricket Team, (who happened to be at the hotel,) and was christened 'The Mugger'! Well done, Allan and well done, Louise. Thank you and our grateful thanks to all the sponsors.

To all who helped to make the weekend a success and gave a boost to Society funds…THANK YOU!

Anne Crotty

We hope you have enjoyed this conference report, don't forget, all of the contact details can be found at the beginning.

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