

# Inside Moebius; ESPRA 2 Copenhagen Jonathan Cole.

Introduction.

Moebius Syndrome or sequence since it is not a singular condition – congenital abnormality of development of the rhomboencephalon. Its cardinal features are;

Abnormality cranial nerves	VI and VII
Horizontal gaze problems in	48%
Abnormality cranial nerve	V 16%
	VIII 29%

Dysphagia

Language development delay	55%
Autism and learning difficulties	5 - 50%

Hand problems	61%
Foot problems	69%

Clumsiness	88%
Poor coordination	83%
Late motor milestones	68%
Hypotonic	31%

i.e. long tracts involved.

Genetic problem likely to affect HOXA1 genes, Midbrain pathfinding and crossing abnormalities ROBO3.

Narratives;

1. Beginnings.

One mother told me;

Sian's father was in the army and she was born during a posting in Germany. Living in a camp, away from many old friends and family, her mother, Mary, struggled, not least because they were avoided by the other Mums and families; Mary grieved for the loss of the child she had wanted. Looking back, if there had been a pre-natal test she would have considered termination. She found it so difficult that Sian could not show any facial response to what was going on around her, whether a noise, or a light or to her – not a smile, not a grimace, not

one of the funny faces most babies make. It was frustrating, lonely - and maddening; this little thing she loved so much didn't respond. At times she wanted to hit her just to get something back. How could you know if she was happy, sad, uncomfortable or bored? How was she to know what her baby was thinking or feeling? How could she care for her and love her if she couldn't read her?

As she grew so Sian proved the doctors wrong. She crawled and laughed and showed her family that she wasn't a vegetable after all. When they were finally given the diagnosis of Moebius, when Sian was around 3, it came with a relief. Moebius Syndrome was a fairly minor thing compared with what they feared might be wrong.

Another mother told me;

'I was 39 and had waited so long for this. The name [of the syndrome] was not important, he was my much loved baby. I talked to him all the time; the nurses and doctors hid behind the curtains checking, listening and then went away satisfied. No smile - he was still my baby; some brain damage, still my baby.<sup>5</sup> They told me that his lack of a smile might lead to problems with bonding. I never could understand that crap.'

'He looked into my eyes. I had no clue that babies move their faces; He moved his arms and legs, so? I talked to him all the time.' Weeks later she went round to a friend's house to see her baby and was slightly surprised. 'I saw this friend's baby doing all these things with his face and I thought "Ugh, that's too much." My friend just smiled and said that is what they are supposed to do.'

Discuss Meltzoff and Moore's work on early neonate imitation. Cross modal sensorymotor integration, a seen smile or whatever translated into a motor act which in turn is felt through proprioception. What internal emotional experience goes with this is unclear, though most parents would say deep and intense.

But few ask why such an important and complex development has occurred; many mammals do little actively for the first few days. Social interaction, showing someone is home, is obviously crucial.

## 2. Martha.

Feeding problems sent her to the special care baby unit. By next morning she had had a brain scan and other tests, and Martha's parents were given a provisional diagnosis of Moebius Syndrome. The doctors explained about the facial paralysis, that her eye movements were also paralysed and that she might also

need speech therapy. The next day the regional neurologist and eye specialist saw her and a multi-disciplinary support team was arranged.

Partial cleft palate  
Glue ear  
Hearing loss  
Micrognathia  
Squint corrections  
Poor vision

Speech production difficult 1;1 tuition  
Pharyngoplasty to close the pharynx to allow more speech

At school she has the Head and her team, a Special Needs Adviser, a Speech Therapist, and a Teacher for the Hearing Impaired, she coped well. Mum kept an active eye on everything from home.

Games poor with balance etc  
School work also difficult – board to book let alone reading, without lateral gaze some get dizzy reading.

Dental abnormalities  
Foot and hand surgery – Achilles lengthening, tendon release etc.

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Imagine poor vision, and lack of movement of the eyes, so you had to move the head the whole time to see, no balance and clumsy at games, drooling, difficulties in eating and drinking, especially in public, maybe pain making life a trial at the best of times,

Learning difficulties are common, but more in younger people with Moebius and not surprising... as Padberg's group from Holland suggest. 'Mask-like face, strabismus, drooling and speech difficulties contribute to the assumption of mental retardation.'

Verzijl HTFM, Van Es N, Berger HJC, Padberg G and van Spaendonck KPM. Cognitive evaluation in adults patients with Moebius Syndrome. Neurology, 2005, 252, 202-207.

Aged 17-55 mean 29.

Mean IQ did not differ from Dutch population and nor did selective attention and memory.

Autism.

Childhood Autism Rating Scale.

1. Relating to People
2. Body Use
3. Adaptation to change
4. Listening response
5. Verbal communication.

Though some studies have suggested Moebius has an excess of autism and MR, Dutch studies in elder subjects have not found this and indeed suggest autism at 5% is linked to MR not Moebius.

Toronto group still finding high prevalence of autism in young children with Moebius, early on they may have...

But the previous work may have been due to the embodiment which Moebius produces and lack of stimulation etc due to the poor eye sight, poor hearing, impassive, drooling children presented. But also the learning delays due to Moebius of themselves make an early diagnosis of autistic features unsurprising. Somatic straightjacket.

3. Cartesian Child.

Celia;

'I did not do ballet, horse riding etc, I did hospitals and operations. I had the eye doctor and the foot doctor and a speech therapist, who I don't remember, and a face doctor.'

She was never aware of not seeing before these ops but then, as she says, she cannot see well even now.

'My limitations were a fact of life. Not being able to see the blackboard, or not being able to see someone over there. I have, or had, a squint and astigmatism. The shape of the Moebius eye is also different and I cannot move the eyes or move the head so easily, my muscle being not so well developed. Crossing a road is still difficult. I cannot judge when a car over there is going to get to me. I

cannot measure distance and moving, the velocity or whatever is out. As a child I could never catch a ball.'

As well as having the talipes, her feet were also painful. She never told anyone.

'No one asked. When I was 7 I stopped walking because the feet were so bad and I had to go to school in a wheelchair. I don't remember learning to walk. After some surgery I could walk but I never told anyone I was in pain. People don't ask little children. I always remember that as an adult I have had pain, but I don't remember pain as a child.'<sup>1</sup>

The myriad of conditions - feet, mouth, eyes, skin (and other ailments too) - and the countless visits to doctors and therapists had an unusual effect on the way Celia viewed herself.

'I never thought I was a person; I used to think I was a collection of bits. I thought I had all these different doctors to look after all the different bits. At half term other children would go off camping or swimming courses, I would see the doctors, this one, then that one. 'Celia' was not there; that was a name people called the collection of bits.

'I did not like my feet; I liked my spirit because I was strong as a child. I liked my brain; I knew I had a brain. I loved reading and read very early on. I liked that bit. I could think and dream and imagine. I had an IQ test which was very high. I was bright, so I didn't worry about the rest. Even though I was a collection of bits I always knew there was something strong inside that I had a mental dialogue with, but it was not the physical body; it was very separate from the physical.'

In his famous *Meditations*, Descartes, exploring certainty and the nature of identity, doubted his embodiment but could not conceive of existence without mind; this was where his famous dictum, 'I think therefore I am' originated. Celia here seems to be making a similar disjunction between herself as whole, thinking being and her imperfect body; Celia was a Cartesian child.<sup>2</sup> Not only this, she was unable to share anything of her internal, thinking world with others. Most talked down to her, as to other children, as she clocked everything and communicated little. She would have an internal dialogue with herself - with her Celia - in her thoughts and imagination. In contrast her speech with others was about matters of fact. No one seemed to know or care about her situation.

'At 5 the only talking I could do was big, about operations, say to doctors; I could only talk to adults, about my bits, not about me. I could also talk about books. Adults were my friends, not children. I just could not do playing with the other kids. Then, with adults, I would have a conversation but with children I

was a bystander. Children had another language, a word language, a body language, a facial language. They run around and jump up and down and I could not do that because my legs did not work and because of my lack of balance.'

#### 4. The Spectator.

A priest in his 50's who had only been given the diagnosis of Moebius recently, and only then been to a Moebius Support Group meeting, to see others. I asked him how he viewed his face and his self in those days. He answered tangentially

'I have a notion which has stayed with me over much of my life - that it is possible to live in your head, entirely in my head. Whether that came out of my facial problem I don't know. I was very introspective. I divided people into two categories: those who didn't want to have anything to do with me for various reasons and those who did.

When he met his future wife,

'I think initially I was thinking I was in love with her. It was some time later when I realised that I really felt in love.'

'I think there's a lot of dissociation. But I think I get trapped in my mind or my head. I sort of *think* happy or I *think* sad, not really saying or recognising actually feeling happy or feeling sad. Perhaps I have had a difficulty in recognising that which I'm putting a name to is not a thought at all but it is a feeling, maybe I have to intellectualise mood. I have to say this thought is a happy thought and therefore I am happy.

'I think also that I have a fear of being out of control with emotions, feeling something that I can't manage. I have also found it very difficult to communicate feelings throughout my life, whether as a child or with my wife, though I think I am getting better at it now. I don't really know how I communicate happiness or sadness. That's a very hard question. Some people cry when they're sad. I don't. I sometimes felt that I would like to be able to cry but you see I am not really able to cry, my tears can come but there's nothing else. My tears only flow when I eat. I am afraid of such feelings. I try and shut them off.'

'Of course, since I have never been able to move the face, I've never associated movement of the face with feeling of an emotion.'

James-Lange theory of emotion and Damasio's somatic marker hypothesis

suggest that embodied emotional expression necessary, I am happy because I smile, not the other way round.

5. Emotional catch up.

Little boy, 'Why can't I be happy.'

Celia:

'I did not express emotion. I am not sure that I felt emotion, as a defined concept. At my birthday parties I did not get excited. There were people around excited, but I followed what they did.'

She was also preoccupied with whether people would come, so she could be 'one of them,' even though she knew she wasn't.

'I don't think I was happy, or even had the concept of, happiness as a child. I was saddened by being in pain or having horrid things like a blood test. Sometimes I would cry but even that would almost be a delayed reaction. I would have been sad so long the tears would come as I did not know what to do.'

Occasionally tears would come as a way to show people how sad she was but this was often because she could not deal with a situation. Once, a 'helpful' mum at a birthday party told her to close her mouth when eating. She did not know what to say, since she knew already that the mother would not understand. So she cried so she could leave the situation and the party.

At the time I just endured it. I could not express my feelings. Sure, I saw children in the playground laughing, but I always related it back to the physical. I did not know about an emotional world. I thought it might be related to my legs.'

'I knew that being happy was something I couldn't do. Everyone told me I couldn't smile. I never got excited at Christmas. I watched others being excited. I verbalised it but not in an emotional way. I knew things were not as they should have been, even though I did know how they could have been different. I was the eternal happy ending girl. Even though things were pretty grim, as a child you don't have a choice, you cannot just stop.<sup>3</sup> I did not realise I was unhappy, I just was.'

Perhaps this is a one off, but there again maybe not. I went to see Duncan and his mother. He was 10 or so, so moved outside to play - on his own.

'My other children had been precocious; smiling, sitting, standing earlier than

most other children. With Duncan it was quite the reverse... there were odd things; for instance the other children recognised me when I went in the room - Duncan's never done that.

'He was a very cuddly baby but he never returned the smiles. The first time I really understood what was going on inside his head was one day when I was looking for a nappy pin for my youngest and he said, "Me get it", and he went and fetched it from the draw. He was three then. Up until then no-one had known what intelligence Duncan might have.'

'I remember his fifth birthday party he was sat in his high chair and went to sleep; it was just like another day for him. He didn't want to know, he didn't want to play. He doesn't really get excited on birthdays, even his own.

'It is difficult to know when he's having fun. When he comes home from school we don't know how he's feeling, we have to ask him. Everything is questions and answers. He has always been a very placid child. He never really gets angry, never really appears upset.

'One thing, however, that clearly upset him is a lack of friends at school. He would come in from school and go upstairs to sulk and only reemerge when he feels better.'

'I wish I had taken more photographs. Because he never did anything and you usually take milestones, I never took them. He always sits back and listens and stores things for later, much more reflective. We always cuddle him but it's true that probably because he's so thoughtful and reflective our approach to him is less spontaneous. I used to cuddle him but he never really cuddled back. Now I still cuddle him because he's my baby but he just sits there saying "I'm too old for this now Mum".'

I then went to talk with Eleanor, in her 30's and who had similar problems of emotional experience.

'Sometimes, if you are in one state of mind, you need others to know. Emotion came for me when I played the piano. We always had a piano and I had lessons from aged 6. By 13 I was quite competent and I found that my fingers unleashed emotion and expression in me, even though I did not know what they were. I would play one piece again and again in various ways; happy, sad, cheeky, all jumbled up inside.



'Musical notes and pattern imposed a mood... though not always the mood I felt. I might have been in one mood, but another would come out through my fingers, there were channels of all sorts of different things inside me.'

She seemed to be expressing and exploring moods through music before she knew what these moods were. It was as though an artist might have started off with a palette with just grey and then, suddenly, red, blue, green are there to play with. As she played with them so she began to experience them too. It was by playing with the colours that she realised what they meant. Eleanor agreed.

'Yes, I had to learn the palette without the feeling initially and then map feeling on. I grew up with music and heard different tones, even though I was not fully aware of emotions. Since I did not have the language, or the words, for feelings, the music and my fingers would convey them. Often what was conveyed was real pain. They could really say it. At that time, everything was in the fingers. I had no body language.'

It was at university that she developed. For the first time it seemed everything was not defined by her face, for the first time people were all nice to her.

'Over the months I bought clothes like everyone else to wear, hippie clothes, and I started to develop a character. It was maybe artificial, but I could design my own. Most people's evolve as they grow up; mine I picked.

'I wanted to be someone who people would like. Before people had not liked me, so I wanted to be sweet, gentle and likeable. I did not want to be radical – at the time there were lots like that. I did not want to stand out. I just wanted to be non offensive, reliable and so that is what I became. That sort of person meant that I did attract people towards me. By the time I left university I was renowned for knowing everyone and everyone knowing me. How did I go from that first night, not knowing how to interact, to that?

'I learnt body language, interaction, to be comfortable with myself.<sup>6</sup> I think I developed a broad circle of friends, because I found I could express different things with different people.'

There she aped and imitated people around her and became social and a being. But it was not entirely clear that she had learnt to embrace feeling within herself.

Lydia.

'I do not think I had emotion when I was a child but now I have it. How did I get it? It was in Spain. I learnt Spanish in two months but – more – they are very

graphic in their emotional expression. The body language I had learnt and used at university could be exaggerated in Spain, using the whole body to express one's feelings. Over here in England it would be over the top, but there it was fine and because of this I learnt to feel within me.

'At Oxford I had learnt a lot of imitating and mirroring and copying but had not, to a very profound depth, had the feeling. I had been using it to conform and because if I did it I got the response, but I, myself, wasn't feeling. But in Spain everyone is so dramatic. If something awful happens then the world is coming to the end, and if fine you party all night. If sad you burst into tears and then go off to the pub. I had gathered all these skills in language and gesture and then in Spain I could just be me.

'Because of the cultural up regulation of feeling in gesture I learnt to feel. I am not sure how I mapped gesture and feeling onto my body, but I was starting to feel then. I could feel really ecstatic, happy, for the first time ever.'

Darwin said that an emotional feeling can either be expressed, continued and become exaggerated or, if not expressed, be reduced and lost. It is as though it has to be in the continual present, continually expressed to be continually experienced.

'Before, without the expression, I had found the feeling difficult. Once in Spain I certainly had the means, the channel and the vehicle, and the feeling. Before, my thought was frigid or cold. I needed the continuation of a thought into real time expression within the body.'

'I was an intellectual at university. In Spain I experienced emotion. As a child I used to play a musical instrument with emotional expression, but I could not let it out. Now, once I could express there was no stopping me.'

Lydia's new experiences were not of course within and about her alone. They emerged within a rich social, cultural world. In a place where emotions were communicated publicly more than in the UK she learnt, somehow, to experience and as well as imitate feelings.

'When you live and share emotion together then you all experience it together. I met a man with Moebius who lived in Sweden and he was one of the saddest people. I met him in Italy; I hope he learnt something from them. If I had not lived in Spain I don't know how I would have turned out.

90% of speech is normally accompanied by gesture, and this seems reduced in Moebius.

Why reduced emotion.

1. Constitutional reduced emotion related to primary condition.

Some have suggested this might be the case, with neurotransmitter problems common to brain stem and emotional brain areas.

2. Primary alexithymia – reduced emotional expression and experience due to associated problem.

**What is alexithymia?**

In brief, alexithymia is *the inability to talk about feelings due to a lack of emotional awareness*. Alexithymics are typically unable to identify, understand or describe their own emotions, and the construct of alexithymia refers to some of the chief manifestations of this deficit in emotional functioning.

**What are the key features of the alexithymia syndrome?**

Difficulty identifying different types of feelings  
Difficulty distinguishing between emotional feelings and bodily feelings  
Limited understanding of what caused the feelings  
Difficulty verbalising feelings  
Limited emotional content in the imagination  
Functional style of thinking

For Freyberger, *primary alexithymia* is an enduring feature of a patient's profile, like a personality trait, that changes little over time or with changing circumstances. *Secondary alexithymia*, on the other hand, occurs in reaction to severe psychological trauma, whereby a patient suppresses painful emotions as a temporary defence against trauma; when the psychological stressor is removed, the alexithymia disappears. Lack of enjoyment and pleasure-seeking  
Stiff, wooden posture

3. FFH bodily feeling of emotion required and without the face this is defective... James Lange Theory of emotion, 'Why Can't I be Happy?' Without face and gesture... it is not elaborated.

4. Social induced model... without reinforcement by others don't learn expression of emotions. Internalisation from Vygotsky and language and social model

- a. express socially and learn shared understanding

- b. internalize and experience oneself.

Somatically induced alexithymia... showing the importance of facial expression for development of all...

Unknown and may well be multifactorial, but the latter two seem important from the narratives. Mother-child interactions obviously important and mirroring of face and gesture.

The portrait.

Set up in Berlin... morning and afternoon.

I need desperately to project myself to show the real me to the world and help the spectator form his opinion and view of me accurately and not based on confusion or misinterpretation.'

Then we brought the portrait from the morning out for comparison. All were agreed that the second was more relaxed, with less nervousness on the face. In the afternoon, evidently, she was more at home with the situation and Alfredo had shown this. Henrietta was puzzled,

'This begs the questions how did he know, how could he sense that - with no words, and no gesture or expression being expressed?'

I suggested that people need to look at the face but also beyond it to decide what a person was like, since only a few facial expressions were unambiguously shown on the face. I then suggested that Henrietta, having Moebius, was actually unable to move her face at all. A photograph in the morning and in the afternoon would not have shown any difference, except for lighting, and that all the differences, in emotion and feeling and situation therefore could only have come from Alfredo's process of portrayal and the audience's subsequent interpretation, and that it was very difficult to know how correct these were. Henrietta agreed.

'I absolutely - hugely - endorse this. The audience could not possibly be able to see physical feeling or emotion. The portrait was open to the artist's interpretation and the audience's take on it. How could they be accurate? Inaccuracies might reflect my mis-communication and their lack of comprehension of me.'

Henrietta had been surprised and slightly shocked by the portraits. She had always thought, and been told, that she could not express anything on her face, let alone subtle emotions. After all, she had no movement of the muscles of facial expression at all; people with Moebius do not have one fixed expression, as in a

photograph or portrait, but almost an absence of expression. He had painted a series of people with dementia; in those relatively early he saw confusion, 'What is next, who I am, where will I go?' But when severe, 'It was terrifying to interpret no feeling.' In was impossible to see Henrietta with no feeling, but equally very difficult to place that feeling on her face, for her or for Alfredo.

Yet in Berlin the audience had agreed on the expression and emotions in Alfredo's two portraits. Either the audience had projected these feelings onto the painting, or Alfredo had successfully introduced them, somehow, into his portrait, whether or not Henrietta felt them. Either way, Henrietta had much to think about. Perhaps people are so used to interpreting facial expression that they create expressions on faces even when they are not there, just like they see a man in the moon.

'For a person with Moebius, where the potential for miscomprehension or labeling is high, the necessity to convey one's self accurately is enormous. It is scary for me to be so open to interpretation [from my face] with so much scope for wrong analysis.'

We had anticipated that, by showing Henrietta's immobile face, we could stress how important it was to go beyond the face to build up a picture of another and so to subvert the genre of portraiture and of image. Instead we had been amazed at the agreement within the audience about the first two portraits. Alfredo and the audiences seemed to have found meaning on the face where none was present, seamlessly taking from the context and situation feelings which were then projected onto the face. Even when we pointed out that she could not show emotion on the face, it seemed to make little difference to them. Henrietta found this very difficult.

'I cannot put my best face on. I have no control. Someone else may put on that most beautiful or sexy face. I cannot do this. In communication I can control a conversation, but I cannot control anything in the portrait or through my face. It is all up to Alfredo. Were these differences there at the time of painting or did they come later? I seemed to be left with the unanswered question; was this emotion really there, did it really exist? It is so important for me that people understand me and don't form a false impression.'

Alfredo listened and then, out of the blue, said,

'As a spectator, what is most difficult for her to express is her consciousness.'<sup>4</sup>

Henrietta went away trying to work out how she could control interactions with others through her face, as well as through gesture and speech, to avoid others

interpreting what they saw. After a lifetime of thinking that people took nothing from her face, and all from her intonation and gesture, she had much to reconsider.

Conclusions.

Shaun Gallagher's book has as its theme how the body shapes the mind. Here we have the ways in which the face shapes the mind. How emotion may depend on embodied expression of it and as importantly the social expectations and how my body and my mind shapes yours.

Merleau-Ponty I exist in the facial expression of the other...

Wittgenstein - I see consciousness in the face...

Without a mobile expressive face our perceptions of others must be changed and we really need not a suggestion of facial equality, like racial equality, but of a recognition that the normal can be expanded.

**All taken from the forthcoming book;**

**Jonathan Cole with Henrietta Spalding, 'The Invisible Smile; living without facial expression.' OUP.**