fx info

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Winter 2013

support@fragilex.org.au
www.fragilex.org.au

In this edition of *fx info*

Design a Fragile X Christmas card!

Christmas Card Design Competition in July

Each year Fragile X Christmas cards are made available for sale to members and to the general community. The aim is to raise awareness of FX and raise funds to support the services provided to members.

This year we'll create unique Christmas cards based on a design submitted by one of the children.

The winning designs will be used on the cover of the Fragile X Association Christmas cards for 2013. Winners will also receive a t-shirt printed with their winning design.

Competition open until 1 August. *See entry form on back page*.

Association News

July is an ideal opportunity for raising community awareness of Fragile X as **July 22** is national **Fragile X Awareness Day**. If you, your family or friends would like to promote awareness of FXS in your community, let us know if we can help – we have posters, lapel ribbons and t-shirts; we also have pens available for sale. *support@fragilex.org.au* or freecall *1300 394 636*.

I'm delighted to confirm that the series of **Family & Community Days, Workshops & Clinics** is going ahead as planned in **September & October**, taking place in Adelaide, Brisbane, Launceston and Sydney.

The **Family & Community Days** will provide a valuable opportunity for families, carers and health & support professionals to learn more about Fragile X syndrome and be updated on latest developments. We're grateful to Professors Randi and Paul Hagerman, and Louise Gane from the M.I.N.D. Institute in the US for once again joining us to lead sessions in Adelaide, Brisbane & Sydney, together with Dr Mike Field, Dr Jonathan Cohen, Professor Kim Cornish, Professor Sylvia Metcalfe and Dr Jan Liebelt. These will be followed by **Children's Clinics** in Adelaide & Brisbane. (see page 5 for details)

Other events coming up include a separate **Adult FX Clinic** presented in Sydney by Dr Jane Law, and a **Workshop and Clinic in Launceston** presented by the Fragile X Alliance team. We are very grateful to the Tasmanian Community Fund for sponsoring the Launceston Workshop & Clinic. (see page 5)

I'm also delighted to announce that in our next newsletter we'll be calling for nominations for the **Fragile X Association Achievement Award for 2013**. Look out for details in the *fx info* Spring edition.

With best wishes,

John O'Connor

President

Running

Congratulations to Torey!

Now in its 22nd year, ***The Sydney Morning Herald Half Marathon*** attracts more than 10,000 participants each May. It's a 21.1km course from Hyde Park, through the city and back. Many runners are sponsored, raising funds for a charity.

This year's race saw well over 10,400 participants. **Torey Clarke** ran to raise funds for the Fragile X Association. Torey is a carrier of Fragile X and has a sibling with the syndrome.

Torey says: *"It was a brilliant experience! Sydney town turned on the weather for the event with a stunning, crisp sunny morning that made the Opera House and Harbour Bridge sparkle. I don't think you could find a more picturesque course. The good news is that I made it in the time of 1 hour, 57 minutes and 38 seconds to be exact*."

Torey raised a fantastic total of $3750! She made it into the top 10 fundraisers of the entire event, coming in at number 10. Torey's husband took some great photos!

On radio and YouTube

Bruce, Rosie, Michaela, Penny

Bruce Donald, FX parent, gave an interview on **RN Drive, on Radio National** in May, profiling the need to raise community awareness of the syndrome.
http://www.abc.net.au/radionational/programs/drive/fragile-x-syndrome/4682724

Bruce's daughter **Rosie** is a keen vlogger and shares her thoughts on the challenges of living with FX and making the most of life on YouTube. *Follow this link and Like!*

http://www.youtube.com/watch?v=Vl9iFCRZaAU

Charity Fun Run & Walk

In Sydney next month **Daniel Wright, Charlie Roberts** (son of FX counsellor, Janie) and **Elliott Green** will be running to raise awareness & funds in the Sydney **Pub2Pub** charity fun run / walk from Dee Why Beach to the Newport Arms: Sunday 25 August.

Pub2Pub
Charity Fun Run & Walk

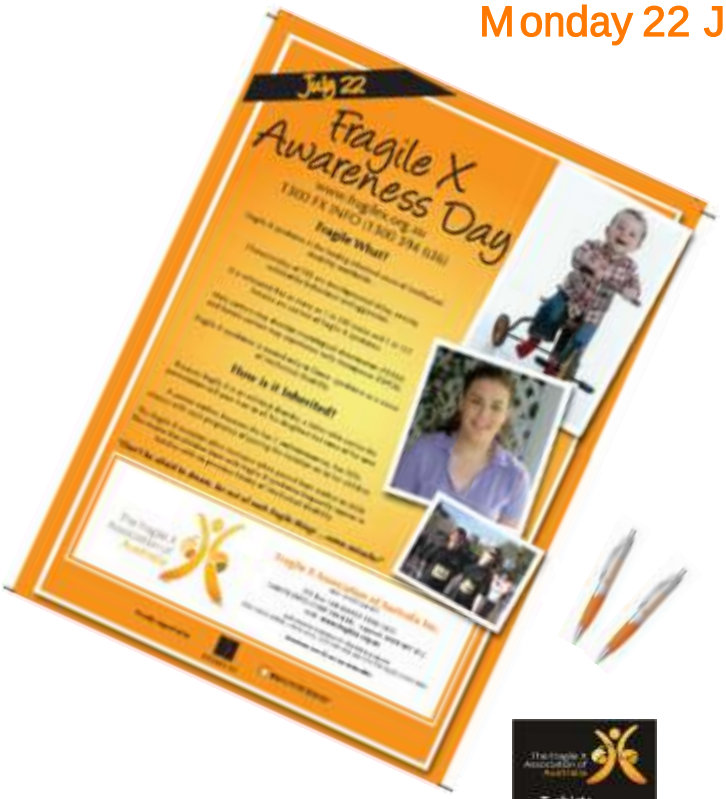
Online Sponsorship for Events Across Australia

gofundraise
Making a difference is easy!

everyday hero

Being sponsored in an event is a great way to raise awareness of Fragile X. The Fragile X Association is registered with 2 online fundraising platforms, so it's easy for you, family or friends to raise funds through sponsorship.
Many walks, runs and marathons in each state are listed on **gofundraise** or **everyday hero,** including: The Age Run Melbourne; City to Surf events in Sydney, Perth, and NT; Bridge to Brisbane; the Great Ocean Walk; Sydney Tower Stair Challenge; Canberra Fun Run; and many other events, large & small.

Monday 22 July: Fragile X Awareness Day



This is the perfect time to raise awareness of Fragile X in your community!

It could be putting up some posters, hosting a morning tea, doing some fundraising, getting sponsored in an event, or running a garage sale!

If you're planning an activity, let us know how we can help.

We have:

- **lapel ribbons**
- **posters** (printed and soft copy)
- **t-shirts** (black with FX logo - limited quantity)
- **pens** - which could be sold for $2 each
- **brochures**
- **donation tins**

Contact us in the office:
support@fragilex.org.au or **1300 FX INFO**

WA community

A **lunch meeting** with the families affected by Fragile X was held on June 22nd at Sienna's Italian Restaurant in Leederville, in Perth, so that the families could finally meet in person.

And Shante also reports that some **morning teas** and brunches are planned for FX Awareness Day in WA.

Also in WA, Jeanie and her sisters are organising a **garage sale** and a local **door knock** in July to raise funds for Fragile X, and have organised for one of the local supermarkets to support the event by providing muffins etc.

Sydney

Sharon Rasmussen and **Simone Zaia** , together with some volunteers will profile FX at the **Sydney Airport Qantas Domestic Terminal** on July 22nd.

They'll be there super early to catch the 6:30am crowds, and will have posters and brochures to hand out to passengers. Sharon has also written a story for the Qantas crew magazine, including the story of her family's experience with Fragile X.

Let us know if you, your family or friends have taken part in or planned an event.
We'd love to include event news and photos in our next newsletter.
Support@fragilex.org.au

Narelle's Story

Narelle Robertson *has kindly agreed to tell us how Fragile X has influenced her and how she has been able to take on new challenges and grow in confidence over the last year.*

She recently represented the Fragile X Association at the launch of the new educational brochures in Victoria where she gave a presentation at St. Catherine's school. Previous to this, in March, she also bravely gave a presentation to her local Lion's Club about the effects of living with Fragile X.

Here is her story...

Narelle Robertson

Please tell us about making a presentation to the members of the Lion's Club and what you said about yourself.
I felt very exposed as I have never admitted to having Fragile X as I wanted to be like everyone else. But I had a wonderful lady I call 'Gran' who is named Barbara who got up with me so it wasn't so bad. I told them the story I had written to the Fragile X Association and how things impact on my life.

You also very kindly agreed to give a presentation at St. Catherine's school on behalf of the Fragile X Association. How was that?
Very scary at first but once I heard the other speakers talk about Fragile X and how they wanted to help, I felt proud and more confident and relaxed.

How would you describe the changes in you over the last year?
By allowing myself to not feel ashamed by having Fragile X and not trying to be something I'm not and having a very supportive family and a bunch of very special friends at the Wonthaggi Lions Club who accepted me for me.

What has made those changes possible?
No more stress and not having to deal with Centrelink. Getting out and joining more things. But nothing would have changed if it had not been for the support of my wonderful mum, Marianne, brother Michael and loving Gran .

Could you describe a typical week in the life of Narelle?
Very hectic! I have genealogy three times a week and garden club every Wednesday. I also do regular Saturday sausage sizzles for my local Lions Club.

What might you say to encourage other people to try out new things?
Don't limit yourself. I did, but once I allowed myself to try new things my life has opened up immensely even if it's only something little at first good things will happen.

Thanks so much for answering these questions. Is there anything else that you would like to add?
Yes, all the staff at the Fragile X Association – thank you so much for all your ongoing support and mostly Janie (when she's not wagging!) I would not have done it without you and I'm very proud to be your VIC Ambassador!

Narelle

Janie Roberts

Feel like a chat? Got a problem and don't know what to do?
Why not phone Janie on the FX support line..... 1300 FX INFO

Janie is the Fragile X Association Counsellor and encourages families, couples, siblings, and individuals to contact her for face to face or phone-based counselling support at our office in Manly, NSW. Counselling offers empathetic listening, a non-judgemental attitude, a safe place to vent, and an opportunity to consider new insights and perspectives.

Janie has experience with counselling in areas of anxiety, depression, isolation, grief and loss, finding balance, feelings of guilt, maintaining hope, relinquishing care and changes in relationships.

Janie has a postgraduate diploma in counselling and psychotherapy. She is a clinical member of Counsellors and Psychotherapists Association of New South Wales Inc (CAPA) and Psychotherapy and Counselling Federation of Australia (PACFA).

Family & Community Days

Randi Hagerman Louise Gane

The Fragile X Association of Australia has arranged for experts from the US and Australia to present an overview of Fragile X Syndrome for families, health professionals and educators in **Adelaide, Brisbane and Sydney** in September.

This is an opportunity to learn more about Fragile X syndrome and be updated on the latest developments. Topics covered will include: **current and future screening options, issues for carriers, available treatments and management of FXS, issues with FXTAS**. There will also be an FXS Discussion Forum.

Speakers will include **Prof Randi Hagerman, Prof Paul Hagerman, Prof Don Bailey, Louise Gane, Dr Jonathan Cohen, Dr Mike Field, Prof Kim Cornish, Prof Sylvia Metcalfe, Dr Jan Liebelt,** and **Michael Cohen**. The speakers are recognised experts in their field and will cover a wide range of management strategies relevant to developmental disabilities and autism.

The keynote speaker will be Prof Randi Hagerman, a Developmental and Behavioural Paediatrician, and the director of the Fragile X Research and Treatment Center at the M.I.N.D. Institute, in Sacramento, California.

A Workshop will also be held in **Launceston** on October 31, presented by the **Fragile X Alliance** team.

Family & Community Days / Workshops	
Adelaide	Monday 16 September
Brisbane	Monday 23 September
Sydney	Wednesday 25 September
Launceston	Friday 1 November

Clinics in Adelaide, Brisbane, Sydney and Launceston

Professor Randi Hagerman and Louise Gane from the M.I.N.D. Institute in Sacramento, California have very generously agreed to offer **six specialist Fragile X clinics for families, following on from the Family & Community Days** in Adelaide and Brisbane. The clinics, with places for children or adults, will be held from 9.00am and will each last half an hour. The clinics will be free for Fragile X Association members.

In addition there will be a regular **Adult Clinic** held in **Sydney** (presented by **Dr Jane Law**), and a **Clinic** in **Launceston**, presented by the **Fragile X Alliance** team. The Workshop & Clinic in Launceston have been generously sponsored by the Tasmanian Community Fund.

Clinics		
Children / Adults	**Adelaide** **Brisbane** **Launceston**	Tuesday 17 September Tuesday 24 September Thursday 31 October
Adults	**Sydney**	Monday 14 October

Please be aware that clinic places are limited, so bookings are essential

For booking information and further details:
Please see the Fragile X Association website for booking details for Family & Community Days, Workshops and Clinics: **www.fragilex.org.au**

Please get in touch with **Joan** if you have any queries:

1300 394 636 (free call) or email to Joan@fragilex.org.au

From the Office….


Janie


Joan


Liz


Wendy

While we each work part-time, there is someone in the office every week day.
If we can help with anything please get in touch:
support@fragilex.org.au
1300 FX INFO

Diary Dates

July 22	**Fragile X Awareness Day**
August	**Children's FX Xmas Card Design Competition** ▪ entries close 1 August ▪ winners notified by 31 August
	Member photos & stories for Spring newsletter: send by 31 Aug.
September	**Family & Community Workshops & Clinics**: **Adelaide** 16/17th, **Brisbane** 23/24th, **Sydney** 25th
	FX Achievement Award 2013 - launched in Spring newsletter
October	**FX Adult Clinic**, Sydney 14th
	FX Clinic & Workshop, Launceston, Oct 31-1 Nov
November	FX Christmas cards available for sale
	FX Achievement Award presentation
	Member photos & stories for Summer newsletter: send by 30 Nov.

Membership Renewals for 2013/2014 due
Liz sent out membership renewal forms in June.

Let us know if you have any queries about renewals:
support@fragilex.org.au

Fragile X Adult Clinic in Sydney: Places Available

We have 2 places available for the **Adult Clinic** in **Sydney** on **Monday 14th October 2013.** The clinic is available for individuals aged 17 years and older.

The initial appointment will be with **Dr Jane Law,** who has extensive experience of Fragile X, at The Developmental Disability Clinic, Royal Rehabilitation Centre in Ryde.

This will include a full medical examination and Dr Law will give advice regarding medications and services that can be accessed by the person with Fragile X. If necessary, referrals can be made to other specialists.

If you would like to secure a place at the Adult Clinic or know of anyone who may benefit from this free clinic please send an email to joan@fragilex.org.au or call the office on 02 9977 0074.

Hi Everyone!
I've just recently joined Janie, Joan and Liz in the office. I've taken over from Linda as Office Manager. I'll be putting together the newsletter each quarter, and I'll also be organising the FX Children's Xmas Card Competition and the FX Achievement Award for 2013. I can help provide information and support to members organising awareness or fundraising events. I'm in the office every day except Thursdays.
Before joining Fragile X Association, I've worked with several small non-profit organisations. Originally from country Victoria, I'm now based in Sydney with my husband and our two primary school-aged boys.
I'm really glad to be working with the Fragile X team and look forward to meeting you over time.
wendy@fragilex.org.au

New Genetic Screening Program Includes Fragile X Test

Genetic carrier testing for fragile X syndrome (FXS) can give individuals and couples information about their risk of having a child with FXS and of developing other fragile X-related disorders. Until recently fragile X carrier testing has only been offered to people who have a family history of FXS or a family history suggestive of FXS or fragile X-related disorders. However, in most families impacted by FXS there is no family history of these conditions.

By Dr Alison Archibald

In Victoria, a new screening program has recently become available which includes genetic testing for FXS. This screening program is run by Victorian Clinical Genetics Services (VCGS). Screening is offered by general practitioners and obstetricians to people planning a pregnancy or in the early stages of pregnancy. Screening is available to anyone interested in testing even if they do not have a family history. The test also includes screening for two other common inherited conditions: cystic fibrosis (CF) and spinal muscular atrophy (SMA).

The screening is being offered because the chance of being a carrier is fairly high (in Australia, 1 in 25 people are carriers of CF, 1 in 40 people are SMA carriers, and about 1 in 150 people are FXS carriers). Additionally, these conditions all have a significant impact on the individual and their family, there are accurate tests available, and there is good research evidence to suggest that screening should be offered.

Screening can be arranged by a general practitioner or obstetrician. For people with a family history of any of the conditions, it is worth contacting VCGS or a clinical genetics service first to discuss testing options. The test is done via a simple blood sample which is sent to VCGS for analysis. Couples who are identified as carriers and have an increased risk of having a child with any of the three conditions are offered genetic counselling.

We hope that by offering this screening more people will have the opportunity to learn whether they have a risk of having a child with these conditions which can then inform reproductive choices (including the possibility of using pre-implantation genetic diagnosis – an IVF technique which involves testing embryos to see if the condition has been passed on).

For further information about the program please contact:
Dr Alison Archibald, Victorian Clinical Genetics Services on 03 8341 6201

Finding Inspiration...

Karin, one of our members, has shared this poem, written by American poet Max Ehrmann in 1927:

Desiderata

Go placidly amid the noise and haste, and remember what peace there may be in silence. As far as possible without surrender be on good terms with all persons. Speak your truth quietly and clearly; and listen to others, even the dull and the ignorant; they too have their story. Avoid loud and aggressive persons, they are vexations to the spirit. If you compare yourself with others, you may become vain and bitter; for always there will be greater and lesser persons than yourself. Enjoy your achievements as well as your plans. Keep interested in your own career, however humble; it is a real possession in the changing fortunes of time. Exercise caution in your business affairs; for the world is full of trickery. But let this not blind you to what virtue there is; many persons strive for high ideals; and everywhere life is full of heroism. Be yourself. Especially, do not feign affection. Neither be cynical about love; for in the face of all aridity and disenchantment it is as perennial as the grass. Take kindly the counsel of the years, gracefully surrendering the things of youth. Nurture strength of spirit to shield you in sudden misfortune. But do not distress yourself with dark imaginings. Many fears are born of fatigue and loneliness. Beyond a wholesome discipline, be gentle with yourself. You are a child of the universe, no less than the trees and the stars; you have a right to be here. And whether or not it is clear to you, no doubt the universe is unfolding as it should. Therefore be at peace with God, whatever you conceive Him to be, and whatever your labors and aspirations, in the noisy confusion of life keep peace with your soul. With all its sham, drudgery, and broken dreams, it is still a beautiful world. Be cheerful. Strive to be happy.

Marty Campbell's Visit to the MIND Institute in Sacramento, California

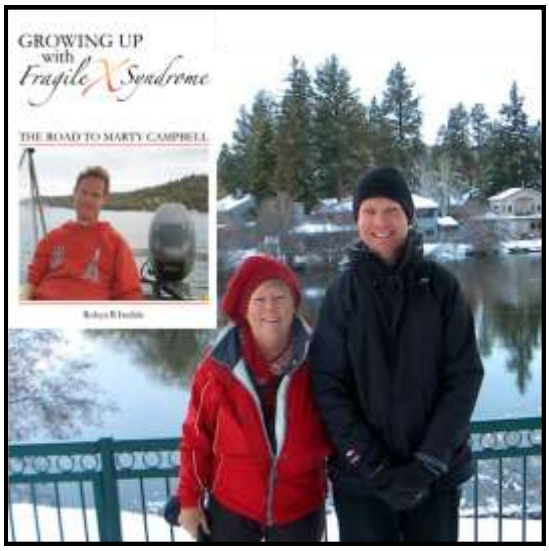
By Robyn Iredale

In my 2011 book about my son Marty, *Growing Up with Fragile X Syndrome: The Road to Marty Campbell*, I described Marty as a pre-mutation carrier with a repeat size of 165. He was an interesting case as he appeared to have more difficulties than one would normally expect of a person with this number of repeats.

For this reason, we were invited to pay a visit to the M.I.N.D. (Medical Investigation into Neurological Disorders) Institute in the US so that researchers there could do a more detailed examination and testing of him.
(**UC Davis M.I.N.D. Institute**: http://www.ucdmc.ucdavis.edu/mindinstitute/research/esdm/index.html)

So just before Xmas 2012, my husband Tim, Marty and I set off for the US. I had arranged a house swap in Bend, Oregon, so that we could include some skiing in our trip. This turned out to be marvellous and we spent some lovely days on Mt Bachelor, which is 22 miles out of Bend. Marty was able to have private ski lessons, under the Oregon Adaptive Sports program, and this brought him on so much that it became very hard to catch up with him!

On 8 January we set off to drive to Sacramento, which was about an 8-hour trip. Marty was a little anxious but we repeated Randi Hagerman's words that 'it would be fun' like a mantra.



Robyn and Marty

Marty had a three day program ahead of him: consisting of neuropsychological assessments, blood and skin samples being taken, psychiatric and behavioural assessments, balance and tremor testing, an MRI scan and a check of his vital signs. He engaged quite happily with all but the neuropsychological assessment, which he tried to avoid by saying that 'he was not doing that baby stuff' (basically IQ testing).

I was also interviewed about Marty and my family's history of FX, which includes my grandfather and mother, two brothers, my children and grandchildren and nieces and their children.

The visit was extremely worthwhile and Marty came away feeling very positive about himself. The staff treated him so nicely that he told them he would be happy to return. In fact he keeps saying to us that people with all sorts of problems (eg drug and gambling addictions, mental health issues) should go to the M.I.N.D. Institute!

The main benefit was that we now have a more accurate diagnosis of his Fragile X status. We now know that Marty's number of repeat sizes ranges from 30 to 500 in different cells, that is he is mosaic. Some cells carry a full mutation and, fortunately, 70% of these full mutation cells are unmethylated alleles. This means that approximately 70% of his full mutation cells have an allele that could be transcriptionally active (i.e. producing the fragile X protein, FMRP). FMRP is necessary for the brain to function normally. It has taken me some time to understand these results but I feel now that we have a much better explanation of his behaviour and level of achievement.

On my return, I contacted the experts in Australia to try to understand the different diagnosis that we now had. It appears that one test that he had in Australia in 1992 showed he was mosaic with 160, 200 and 400 repeats, the last two bands being less prominent. Somehow, I never received this result or I mislaid it!
(… continued on next page)

Marty's Story, *cont'd*

The major outcomes from Marty's trip to the MIND Institute are:

- Marty's increased self image
- Our better understanding of Marty's mosaic FX status and IQ
- Our awareness of the critical importance of methylation (now being analysed by Dr David Godler in Melbourne)
- The dramatically lowered possibility of Marty developing FXTAS, as his MRI scan showed no white matter disease or atrophy of the brain
- An increase in Marty's anxiety-reducing medication and the addition of minocycline to his medications — all working well!
- The recommendation that my non-carrier daughter (who already has two healthy children) be retested to check on the validity of her result of 1992 (*the following article by Dr Mike Field canvasses who should consider being retested)*
- My return to taking medication for depression — having been convinced by Louise Gane that depression kills brain cells and medication actually promotes the development of new brain cells (hopefully warding off Alzheimer's disease).

New Research Study

By Dr Mike Field

Marty's case just shows how different Fragile X testing is today than it was 20 years ago. Diagnosis until recently relied on a type of test known as a Southern blot that is tricky to perform and may not always show the full range of sizes or how active the Fragile X gene is. This was the case for Marty. Newer sizing tests that rely on different technology are much better at detecting the full range of premutation expansions present in the Fragile X gene. Despite more accurate testing, we still see some families where only premutation alleles are detected, but where there are symptoms suggestive of Fragile X syndrome.

To improve testing, we can now see how active the Fragile X gene is in each patient (a test known as methylation). Sometimes patients classified as having only premutations from standard testing may show Fragile X gene inactivation (gene methylation) explaining why they have clinical features of Fragile X syndrome. These testing methods will not change results for family members who have results in the normal (non-carrier) range and who are clinically normal.

Methylation testing is available through Victorian Clinical Genetics Services (VCGS) and can be requested through GOLD Services or VCGS or your local doctor if a Fragile X result from standard testing is not consistent with the patient's symptoms.

In 2013 NHMRC has awarded a 3 year research grant of $650,000 for a study led by A.Prof Howard Slater and Dr David Godler at the Murdoch Children's Research Institute in recognition that more research is required in this area. This multi-centre study will involve patient assessments at: Olga Tennison Autism Research Centre at La Trobe University led by Prof Cheryl Dissanayake; Department of Psychology at the Royal Children's Hospital led by A. Prof Lesley Bretherton; VCGS Clinical Genetics group led by A.Prof David Amor; and GOLD Services conducted under the supervision of Dr Mike Field.

The study will be primarily conducted in Victoria and potentially through sites in NSW with an aim to find out how early after birth these new methylation tests can be used to predict intellectual disabilities, behavioural problems and autism in children and adults who have expansions in the Fragile X gene. The other study aim is to find out why some people with premutation expansions in the gene develop medical problems while others don't. Therefore, there is the potential that this research will lead to earlier diagnosis and a better understanding of the needs of families with premutation and full mutation gene expansions. Hopefully this may lead to improvements in quality of life through earlier access to intervention programs.

Further details of the study will be available in the next newsletter, and may be one way to get further testing if you feel your family member requires more investigation.

In the meantime if you think a family member should be tested using these new methylation tests please call:

NSW:	**GOLD Services**	Ph: (02) 4985 3100	Fax: (02) 4985 3105
Vic / Tas:	**Victorian Clinical Genetics Services (VCGS)**	Ph: (03) 8341 6258	

If you are from other states please contact your local doctor to request methylation testing at VCGS.

Behavior Modification in the Classroom: Braden on Behavior

Marcia L. Braden, PhD is a licensed psychologist and special education consultant in who specializes in Fragile X. *This is the first in a 4-part series from the website www.fragilex.org, reproduced with the permission of the author.*

During a recent school consultation, I was reminded of how the behavior of students with fragile X syndrome is often misunderstood in the classroom. Watching a student with FXS struggle is difficult when his behavior is affected by those characteristics that make up the Fragile X phenotype. (See chart below.) The fact that sensory input is difficult for him to interpret or that his speech production is cluttered and hard to understand or that his learning style is counter to the way teaching is traditionally conveyed may be the very reasons he is acting out or refusing to participate.

An example most parents and professionals familiar with FXS would recognize: The loud and unpredictable sound of the music class next door to our student's classroom causes him to become hyper-aroused, scanning the room waiting for the next sound to be made. After a while, he covers his ears, puts his head down on the desk and screams, unable to manage his anxiety any other way.

Obviously, the student's level of affectedness dictates just how much he can tolerate in his learning environment. His inability to tolerate certain input may result in challenging behaviors. But modifying his classroom behavior requires an understanding of the cause or function the behavior serves. Attempting to reduce the frequency of the behavior without considering its function is an exercise in futility.

In other words, in order to change or modify the behavior, we must understand why it occurs. When we understand, we can be more successful in helping the student with FXS become more adaptable and less disruptive in the classroom.

The idea of modifying behavior is not new. Now referred to as "Applied Behavior Analysis," it simply refers to a variety of strategies to increase or decrease the frequency of certain behaviors believed to enhance or interfere with learning. Many school systems use this term when discussing the treatment of problem behavior with parents. When necessary, a behavior intervention plan (BIP) is written as part of the student's Individual Education Plan (IEP). This plan includes a variety of strategies and supports to assist the student in "modifying" his behavior.

A few words of warning regarding BIPs: When rigid behavior analysis is applied without consideration for characteristics that are part of the FXS phenotype, the plan can be ineffective. In other words, pinpointing the function of the behavior is essential to effective behavior modification. But when the behavior is analyzed in isolation from the FXS phenotype, it may fit the behavioral model (determine the function and apply an intervention), but miss the mark in providing successful intervention.

The following example illustrates the difficulty of applying behavior analysis without considering the FX phenotype. A student with FXS is asked to write his name in class, but he throws the marker and tears up the paper. At first glance, the function of this behavior seems to be a willful attempt to escape or avoid the demands of the task. A behavior intervention plan might employ strategies to support the student to persevere through the task by offering substantial reinforcement such as a desired snack or free time. That's a traditional behavior modification approach, and it works with most children in most settings (with most adults, too, for that matter, though different rewards may apply).

However: This approach misses the point because the escape behavior of the student with FXS may not be willful but merely a reaction to what he experiences as overwhelming anxiety in having to write his name. Many students with FXS have motor planning and executive functioning deficits as well as fine motor delays, making writing extremely difficult. The anxiety and discomfort created by the writing task create a fight-or-flight reaction.

In this example, it is important to go beyond the typical identified function of escape or avoidance, and instead focus on determining the more relevant issue: From what is he trying to escape? When we realize what that is, we know that instead of providing motivation to have the student continue the task, it would be more effective to provide an alternative strategy for him to comply with it. Possible alternatives would be for him to use a stamp to write his name, or to spell it with letter tiles. This might eliminate his need to escape from the writing task.

Ultimately, the request to write would no longer elicit such a negative behavioral response. The student would learn that the expectation was no longer insurmountable and through repeated exposure and appropriate supports such as tracing, writing on a white board with a marker or using a keyboard, he would be more willing to attempt to write. This is the essence of behavior modification.

In another example, a student with FXS becomes anxious whenever a fellow student screams. The screaming is unpredictable and loud. The student with FXS reacts to the discomfort by hitting himself. The behavior is aggressive and could present a significant risk to his welfare. This self-injurious behavior is not premeditated but rather, a reaction fueled by his anxiety. As the analysis is completed, it becomes clear that the only time this behavior occurs is when the other student is present. The mere anticipation of that student's screaming causes the student with FXS to become hyper-aroused and dysregulated.

How to handle this situation? The intervention might include graduated desensitization to the other student who screams. This might be accomplished by allowing the student with FXS to move away from the screaming, to wear headsets to muffle the sound or to leave the classroom to complete a contrived task. This process, however, might be so uncomfortable that the student with FXS continues to hit himself anytime he is in the presence of the other student. In this case, the success of the intervention is contingent on regulating the behavior, not spending time desensitizing him to the screaming by repeated incremental exposure.

Psychologists often use the ABC (Antecedent-Behavior-Consequence) model when analyzing behavior. The consequence actually becomes the intervention. Correct application of the model can mean the difference between successful and unsuccessful intervention. "Antecedent" means whatever was occurring just prior to the negative behavior. When we account for the FXS behavioral phenotype in analyzing the antecedent, the intervention (consequence) will be appropriate and successful. The chart below illustrates the ABC chart with the two different consequences/interventions.

Implementation of the A-B-C Model Without An Understanding of FX Behavioral Phenotype

Antecedent	Behavior	Consequence/Intervention
Writing Task	Tearing paper and throwing marker	Reinforce the student for every minute he perseveres through the task

Implementation of the A-B-C Model With Understanding of the FX Behavioral Phenotype

Antecedent	Behavior	Consequence/Intervention
Writing Task	Tearing paper and throwing marker	Offer alternative writing task, give opportunity to take a sensory break, provide letter stamps, use "backward chaining"*

Backward chaining is a technique that breaks down a task into its sequential steps, with the adult initially doing all but the very last one. When the student is ready, the adult does all but the last two, and so on, until the student has accomplished each step on the "backward chain" and arrives at the beginning

The classroom is a place to learn a variety of behaviors. The student with FXS may need to learn to relate socially, complete academic tasks, demonstrate better speech production or tolerate certain sensory input without demonstrating challenging behaviors. When the intervention does not include an understanding of the behavioral phenotype, but rather requires behavior that is incompatible with what is possible, the intervention will fail. When the intervention fails, teachers and peers may come to view the student as less viable in the classroom, and the situation may needlessly deteriorate.

It is the responsibility of the educators and clinicians to observe the challenging behavior, analyze it, and decide on its function, based on a clear understanding of the characteristics that comprise the behavioral phenotype. This will ensure sound behavioral programming and successful behavior modification.

Characteristics of FXS Behavioral Phenotype
Cognitive deficits:

- Sensory integration dysfunction
- Speech and language delays
- Gross and fine motor delays
- Physical ailments
- Social/psychological deficits
- ADHD, anxiety, depression

Other articles by **Marcia Braden** related to behaviour and children in the classroom on the US National Fragile X Foundation website at **www.fragilex.org** include:

- **Navigating the Road to Inclusion** (February 2013)
- **Reading, Writing & Behavior** (February 2013)
- **Communicating through Behavior** (September 2012)

Design a Fragile X Christmas Card!

The Fragile X Christmas Card competition is open to all children whose families are members of the Fragile X Association.

A **winning design** will be used on the cover of the Fragile X Association Christmas cards for 2013. The winners will receive a t-shirt printed with their own Christmas card design.

Your brief: **Design the cover for a Christmas card for the Fragile X Association**

Some ideas: Your design could be a **drawing** or **painting**, or a **piece of art**.
Or it could be a **photo** you have taken of a Christmas design you have made, or a photo you have taken of something that represents Christmas to you

How to enter: Send your Christmas card design and entry form (below) to us in the Fragile X Association office before **5 pm Thursday 1 August**

Competition Rules:
The Christmas card cover design must be your own original work
The winning designs will be selected by *Janie Roberts*, the Fragile X Counsellor.
Winners will be notified by 31 August and announced in the next edition of the *fx info* newsletter, due out in September.
Competition closes: Thursday 1 August

Any queries:
Contact Wendy at the Fragile X Association office
on freecall 1300 394 636 or email to wendy@fragilex.org.au

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Entry Form: Fragile X Association Children's Christmas Card Competition

Your name: ____

Your age: ____

Your contact details: (phone / email) ______________________________

Your address: ____

note:

Please have an adult sign this to give the Fragile X Association permission to use your Christmas card design on Christmas cards, on the Association website, and in the newsletters:

I (print name) ______________________ **give the Fragile X Association permission to use the Christmas card design by** __________________**on Fragile X Association Christmas cards, website and newsletters.**

Signed:____________________________
Date: ____________________________

Send entry form and design to: Fragile X Association of Australia by **Thursday 1 August 2013:**

Post: to *Fragile X Association, PO Box 109, MANLY, NSW 1655*
or Email: send **.jpg file** to wendy@fragilex.org.au