Challenging Behaviour in Fragile X Syndrome: A Systematic Review and Analysis of the Literature
Rebecca Hardiman & Peter McGill
Tizard Centre, University of Kent, UK

Introduction
Challenging behaviour is an issue of high concern for those caring for individuals with Fragile X syndrome (FXS; Hatton et al, 2002). The aim of this systematic review was to collate available information on the epidemiology and characteristics of these behaviours in individuals with FXS.

Method
5 databases were searched in June 2014 using variations on the terms 'Fragile X syndrome' and 'challenging behaviour'. 525 unique papers were identified, 33 of which were included in the review.

Weighted averages of prevalence were calculated for classes of challenging behaviour in total, as well as individual topographies of these behaviours. Furthermore, the prevalence of different classes of behavioural function was determined. Prevalence differences were compared using Tests for the Comparison of Proportions

Prevalence
- Of those males assessed, 39% engaged in aggressive behaviour and a significantly higher proportion (45%) in self-injurious behaviour (SIB; W=5.15, n=5860, p<.0005).
- A smaller proportion of females showed self-injurious or aggressive behaviours: 14% and 16% respectively.
- Males were significantly more likely than females to engage in self-injury (W=18.43, n=3686, p<.0005) or aggression (W=14.00, n=3497, p<.0005).
- Hand-biting was assessed as a specific topography of SIB. Of those assessed, 44% of males and 15% of females with FXS displayed hand-biting.

Topography
- Biting was significantly the most common form of SIB for males (compared to hitting; W=7.92, n=1235, p<.0005). However, the difference did not reach a level of significance for females. (Figure 1).
- The hand was the most common location toward which SIB was aimed, followed by the head, for both males (78% vs. 47%; W=9.98, n=976, p<.0005) and females (76% vs. 51%; W=3.19, n=102, p<.005).
- The topography of aggressive behaviour has only been researched in males, for whom hitting others was the most common form recorded (67%), followed by grabbing (45%; W=3.09, n=138, p<.005)

Function
- 74% of males' behaviour was at least partly maintained by social-negative reinforcement; significantly more than other classes of reinforcement (compared to social positive (other): W=2.82, n=188, p<.005).
- In Beavers and colleagues’ (2013) systematic review of functional analyses, 32% had escape-maintained behaviour, compared to 59% in our sample.

Discussion
Evidence supports gender-differences in the prevalence and topography of challenging behaviour. Furthermore, the function of challenging behaviour seems to reflect an increased motivation to escape, relative to gaining attention.