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Paraphilic Compulsion Secondary to Dopamine Replacement Therapy and Successful Treatment with GnRH Analogues

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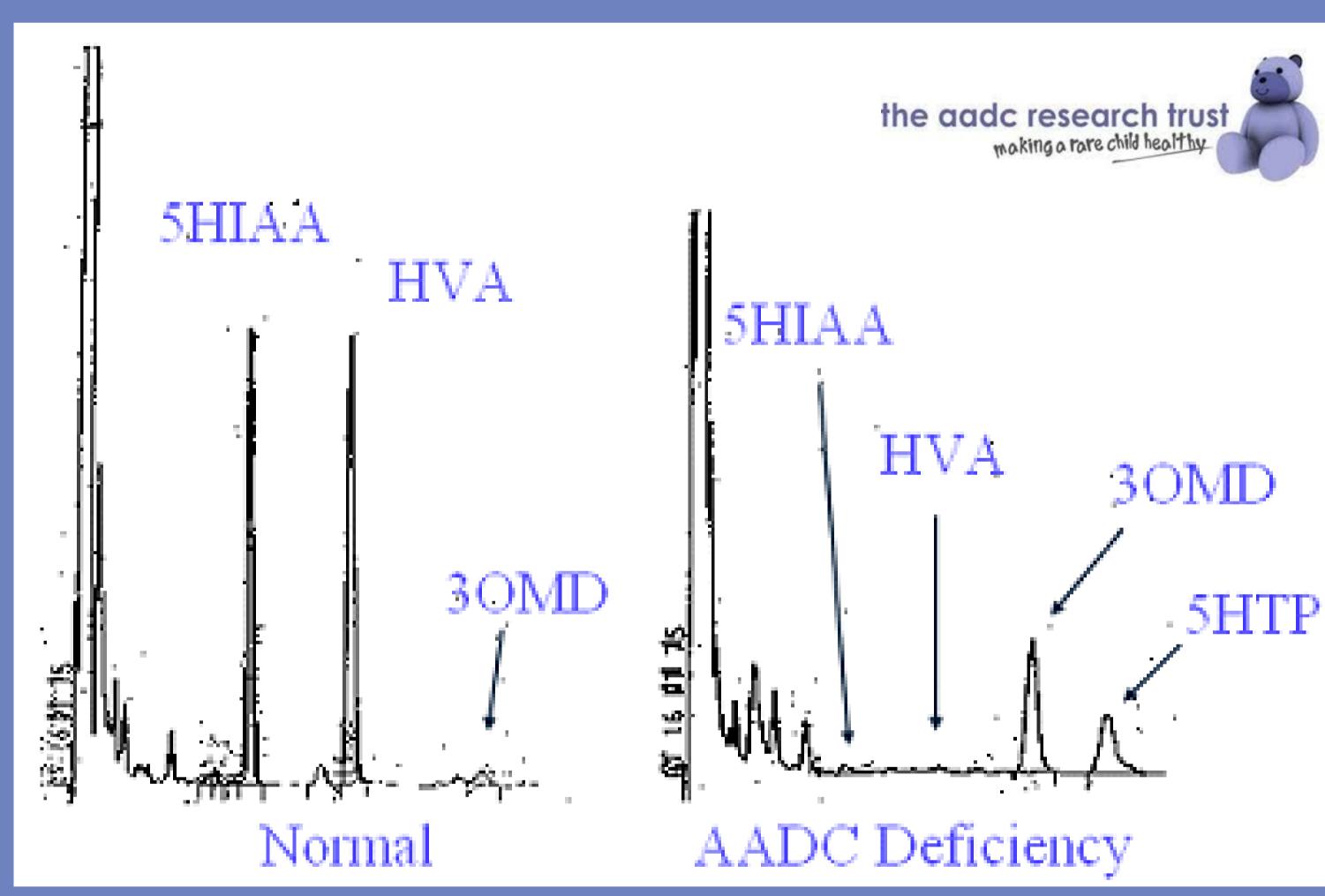
Background:

Hypersexualized behaviour in the paediatric population is a rare phenomenon. The aetiology of paraphilia is not completely understood, but some studies suggest imbalance of the dopamine serotonin system. Paraphilia has also been described as a side-effect of treatment with monoaminooxidase inhibitors (MAOI) and dopamine agonists. Most of the currently used pharmacologic treatments of the paraphilias have serotonin and testosterone/dihydrotestosterone as their targets and treatment options such as selective serotonin reuptake inhibitors (SSRI) and GnRH analogues have been described.

Case report:

We report the case of a 16-year-old boy with background of aromatic L-amino acid decarboxylase (AADC) deficiency, a rare inherited neurometabolic disorder, and autism. AADC is a pyridoxal 5'-phosphate enzyme responsible for the production of the neurotransmitters dopamine and serotonin and is predominantly found in neural and kidney and liver tissue. Its deficiency clinically presents with prominent extrapyramidal and autonomic features and CSF monoamine deficiency with increased 3-O-methyldopa. Management options include use of MAOI and dopamine agonists (DA).

Our patient had been on long-term treatment with tranylcypromine (MAOI) and rotigotine (DA) and developed secondary paraphilia and aggressive behaviour causing severe distress for himself, his parents and carers. After multidisciplinary discussion treatment with GnRH analogues was initiated. This led to persistent resolution of the hypersexualized behaviour and aggressiveness.



High performance liquid chromatography showing characteristic patterns of AADC deficiency.

Conclusion:

This is the first case to describe successful treatment of paraphilia with GnRH analogues in children. It is also the first case to describe paraphilia secondary to MAOI and dopamineagonists in paediatric patients with AADC deficiency.







