Screening for autonomic disorders in ataxia-telangiectasia using the Modified COMPASS-31: preliminary results

E.A.Z.Y. Liew[1], W.P. Whitehouse[2,3]

[1]Paediatrics and [2]Paediatric Neurology, Nottingham University Hospitals NHS Trust; [3]School of Medicine, University of Nottingham; Nottingham UK

Introduction

Ataxia-telangiectasia (A-T) is a rare, multisystem, life-shortening, autosomal recessive disease caused by mutations in the ataxia telangiectasia mutated (*ATM*) gene. In addition to cancer predisposition, and immune deficiency, neurological symptoms predominate including progressive cerebellar ataxia, movement disorders, and sensory and motor neuropathy. Autonomic dysfunction has not been reported in A-T to date.

The Composite Autonomic Symptom Score (COMPASS-31) questionnaire is a robust measure of autonomic symptoms in children and adults.

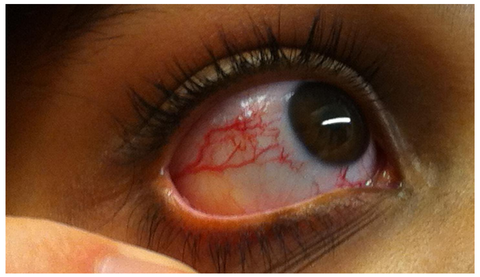
Aims

As we host the UK national clinic for children with A-T, we wanted to pilot a symptom screening tool for autonomic disorders in children with A-T.

Methods

The COMPASS-31[1] and other questionnaires of autonomic symptoms were reviewed, including the Pediatric Autonomic Symptom Scale (PASS), and the Centre for Autonomic Medicine in Pediatrics (CAMP) intake questionnaire. The COMPASS-31 was modified (mCOMPASS-31) for ease of administration and scoring locally.

A convenience sample of children with A-T completed the mCOMPASS-31 together with their parent, at a face-to-face consultation.



With kind permission of patients aged 4 and 17 years and their parents; National Children's A-T Clinic, Nottingham UK.

The scores for each item were altered to represent the weighting for each section, which is normally applied as a section specific factor after questionnaire completion

The scores using the standard method and the new method were compared. Simple descriptive statistics were used.

As the mCOMPASS-31 is a straight-forward brief questionnaire, readily available to clinical teams, no Research ethics Committee review was requested.

Results

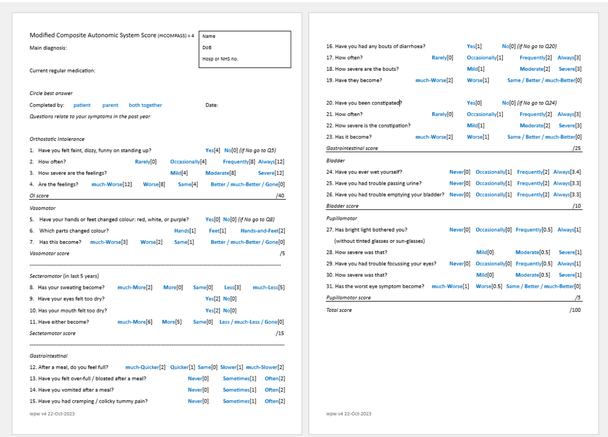
So far 13 children, including 6 male and 7 female, aged 4-16 years (median 11 years 7 months) have completed the mCOMPASS-31.It was quick and easy to administer to children, accompanied and prompted where necessary by a parent. The children with A-T scored 2.0-33.1% (median 7.5; IQR 6.5-11.1) using the standard scoring method. The new scoring system was quicker and easier to calculate but differed by -0.86 to +5.8% (median +1.5). 2/13 (15%) children scored > 20 with both scoring methods.

Discussion

A score >20 is taken by some as evidence for significant autonomic dysfunction. The new scoring system correctly identified these children. However, it tended to slightly over-estimate the score compared to the standard method, which could be clinically misleading.

References

1. Sletten DM, Suarez GA, Low PA, Maandrekar J, Singer W. COMPASS 31: *Mayo Clin Proc.* 2012; 87(12): 1196-1201.



Modified Composite Autonomic System Score (mCOMPASS) v 4

Main diagnosis:

Name
DoB
Hosp or NHS no.

Current regular medication:

Circle best answer

Completed by: patient parent both together Date:

Questions relate to your symptoms in the past year

Orthostatic Intolerance

1. Have you felt faint, dizzy, funny on standing up? Yes[4] No[0] (if No go to Q5)
2. How often? Rarely[0] Occasionally[4] Frequently[8] Always[12]
3. How severe are the feelings? Mild[4] Moderate[8] Severe[12]
4. Are the feelings? much-Worse[12] Worse[8] Same[4] Better / much-Better / Gone[0]

OI score /40

Vasomotor

5. Have your hands or feet changed colour: red, white, or purple? Yes[0] No[0] (if No go to Q8)
6. Which parts changed colour? Hands[1] Feet[1] Hands-and-Feet[2]
7. Has this become? much-Worse[2] Worse[2] Same[1] Better / much-Better / Gone[0]

Vasomotor score /5

Sectiomotor (in last 5 years)

8. Has your sweating become? much-More[2] More[0] Same[0] Less[3] much-Less[5]
9. Have your eyes felt too dry? Yes[2] No[0]
10. Has your mouth felt too dry? Yes[2] No[0]
11. Have either become? much-More[6] More[3] Same[0] Less / much-Less / Gone[0]

Sectiomotor score /15

Gastrointestinal

12. After a meal, do you feel full? much-Quicker[2] Quicker[1] Same[0] Slower[1] much-Slower[2]
13. Have you felt over-full / bloated after a meal? Never[0] Sometimes[1] Often[2]
14. Have you vomited after a meal? Never[0] Sometimes[1] Often[2]
15. Have you had cramping / colicky tummy pain? Never[0] Sometimes[1] Often[2]

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16. Have you had any bouts of diarrhoea? Yes[1] No[0] (if No go to Q20)
17. How often? Rarely[0] Occasionally[1] Frequently[2] Always[3]
18. How severe are the bouts? Mild[1] Moderate[2] Severe[3]
19. Have they become? much-Worse[2] Worse[1] Same / Better / much-Better[0]

20. Have you been constipated? Yes[0] No[0] (if No go to Q24)
21. How often? Rarely[0] Occasionally[1] Frequently[2] Always[3]
22. How severe is the constipation? Mild[1] Moderate[2] Severe[3]
23. Has it become? much-Worse[2] Worse[1] Same / Better / much-Better[0]

Gastrointestinal score /25

Bladder

24. Have you ever wet yourself? Never[0] Occasionally[1] Frequently[2] Always[3.4]
25. Have you had trouble passing urine? Never[0] Occasionally[1] Frequently[2] Always[3.3]
26. Have you had trouble emptying your bladder? Never[0] Occasionally[1] Frequently[2] Always[3.3]

Bladder score /10

Pupillomotor

27. Has bright light bothered you? Never[0] Occasionally[0] Frequently[0.5] Always[1]
(without tinted glasses or sun-glasses)
28. How severe was that? Mild[0] Moderate[0.5] Severe[1]
29. Have you had trouble focussing your eyes? Never[0] Occasionally[0] Frequently[0.5] Always[1]
30. How severe was that? Mild[0] Moderate[0.5] Severe[1]
31. Has the worst eye symptom become? much-Worse[1] Worse[0.5] Same / Better / much-Better[0]

Pupillomotor score /5

Total score /100

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modified COMPASS-31 draft questionnaire

Conclusions

The modified COMPASS-31 is well suited for use with children, however we will need to try it on a larger number of children with A-T, before drawing firm conclusions.

Furthermore, the new scoring system will be adjusted to make the resulting patient scores even closer to the standard scoring results.

Acknowledgements

We are grateful to all the children with A-T and their parents for taking the time to complete the questionnaire.

Nottingham University Hospitals NHS Trust

Nottingham Children's Hospital