

INTRODUCTION

Turner's syndrome (TS) affects approximately 1 in 2500 live female births. Long term management of the condition requires consideration of the multiple systems involved and an annual review "checklist" approach is recommended using a multi-speciality team. We aim to audit current practice of a specialist Turner's clinic in a UK secondary care hospital in order to identify gaps in the current management of these patients.

METHODS

At the time the clinic was established a checklist of recommendations for health screening was developed based on the National Turner's Syndrome Guidelines. For the purposes of audit a check list and gold standards were developed based on the clinic's own recommendations for care. Data for the audit was collected from clinical encounters during the period June 2011-Jan 2013 on a cohort of thirty-seven patients.

RESULTS

Thirty-seven patients attended the Turner's clinic during the allocated time period. Four of these patients were excluded from the audit. Three having moved away from the area and one due to having Noonan Syndrome rather than TS.

Cardiovascular

All patients had their BP measured annually. 89% of these were within target. 61% of the patients had an ECG. 82% of the patients had 3-5 yearly echocardiograms, however all those that didn't had had a cardiac MRI. 85% of the patients have had a cardiac MRI.

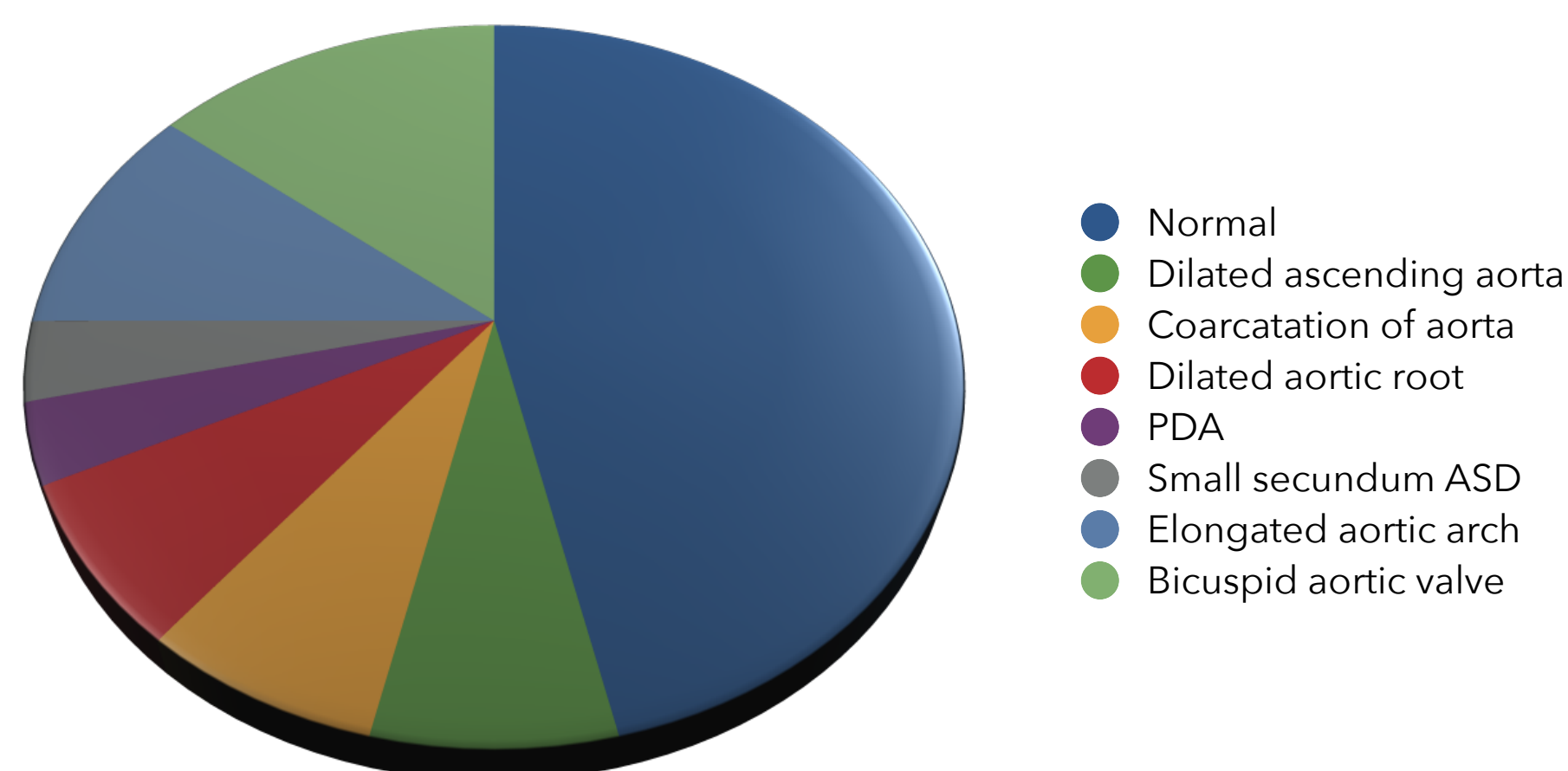
Fertility and hormone replacement

79% of the patients are documented as currently taking hormone replacement therapy. Of these patients, 11.2% are taking a transdermal form with the remainder taking oral medication. Patients not taking HRT are all either >50 years in age, or < 50 years and a mosaic. Therefore 100% of patients that required hormone replacement therapy are receiving it.

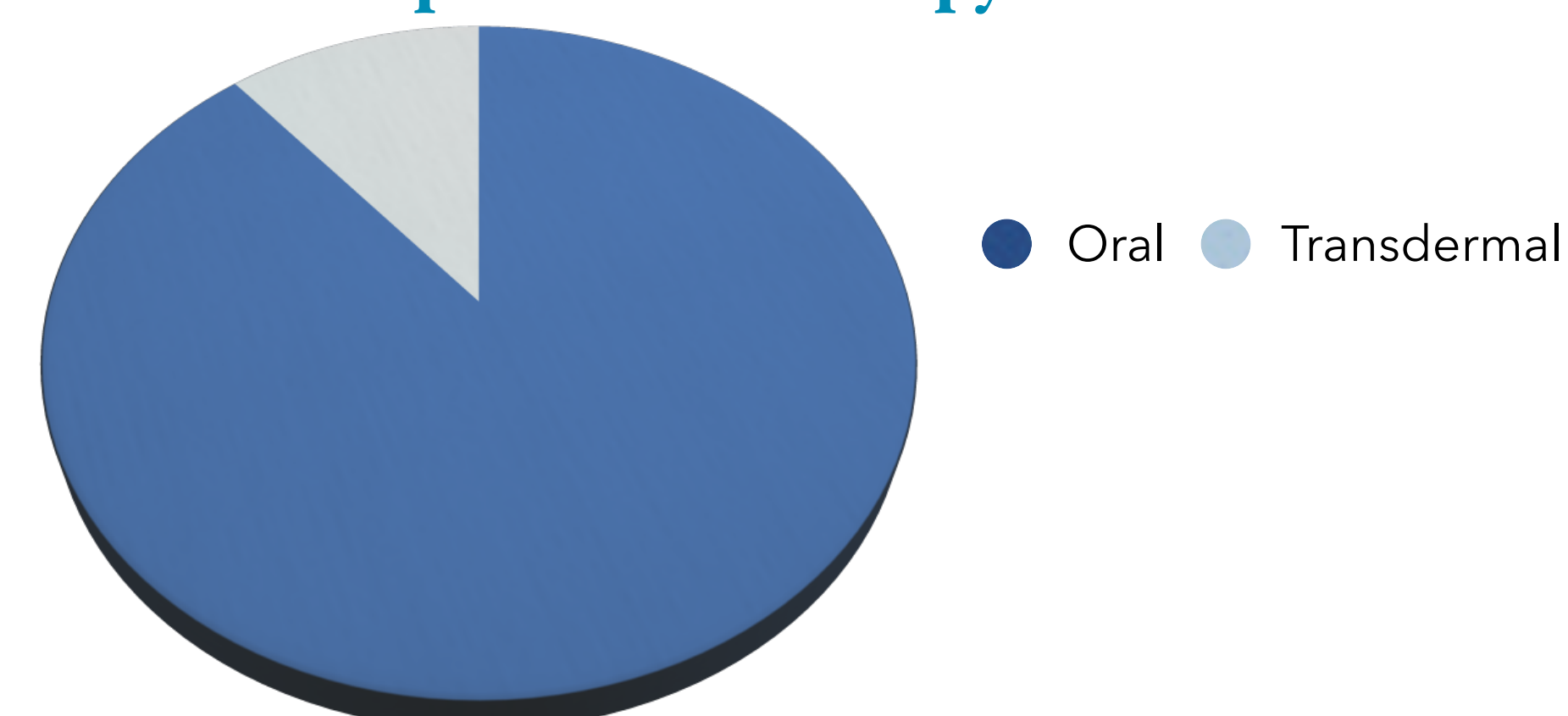
Bone protection

94% of patients had a bone DXA scan performed. 45.1% of the patients had abnormalities on these scans. Out of the patients with abnormal DXA scans aged under 50 100% of them were taking hormone replacement therapy.

Cardiac MRI Results



Hormone Replacement Therapy



DISCUSSION

A standardised multidisciplinary evaluation of Turner's Syndrome patients has been shown to pick up significant previously undiagnosed mortality. ⁱⁱⁱ

Cardiac MRI

Patients with TS have a 6x greater risk of aortic dissection than the general population. Cardiac MRI is the preferred imaging modality for surveillance of cardiac abnormalities in these patients. ^{iv} In this audit MRI picked up abnormalities undetected on echo in 23% of patients.

Hormone Replacement Therapy

The majority of patients were receiving HRT. International guidelines suggest that transdermal preparations may be more physiological. A switch to transdermal was offered in 44% of appropriate patients but less than half of them switched.

Conclusion

This audit has highlighted the importance of a checklist approach to the management of TS patients. The service will continue to ensure:-

1. routinely offering transdermal preparations of oestrogen
2. protocol for initial and repeat cardiac MRI
3. a multi-disciplinary role to management