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BACKGROUND

It is well known that patients with Turner Syndrome (TS) often have prolonged QTc, though the significance of this remains unknown.

Current guidelines recommend TS patients avoid QT-prolonging medication potentially complicating their treatment options.^{1,2,3}

OBJECTIVE

We sought to determine the degree and frequency of QTc prolongation in patients with TS, as well as the effect of medications on QTc and occurrence of any arrhythmia.

METHODS

A retrospective, multi-center chart review was performed of all patients with an ECG and genetically-proven TS.

Medications at the time of ECG were reviewed and recorded; QTc was calculated using Bazette's formula.

Medications were classified by QT-prolonging risk in accordance with www.crediblemeds.com.

Potentially confounding variables were recorded, including karyotype, liver disease, congenital heart disease, bicuspid aortic valve, and age.

Clinical correlates of QTc prolongation were calculated by chi-squared or ANOVA test.

RESULTS

QTc was >440 in 53.4% of patients and >460 in 21.6% of patients

The average QTc of patients on QT-prolonging medications was significantly higher than that of patients not on QT-prolonging medications ($p=0.00002$).

Patients on no medication and non-QT-prolonging medications did not differ in QTc ($p=0.25$).

There was no correlation between QTc >440 or QTc>460 and karyotype, liver disease, congenital heart disease, or bicuspid aortic valve.

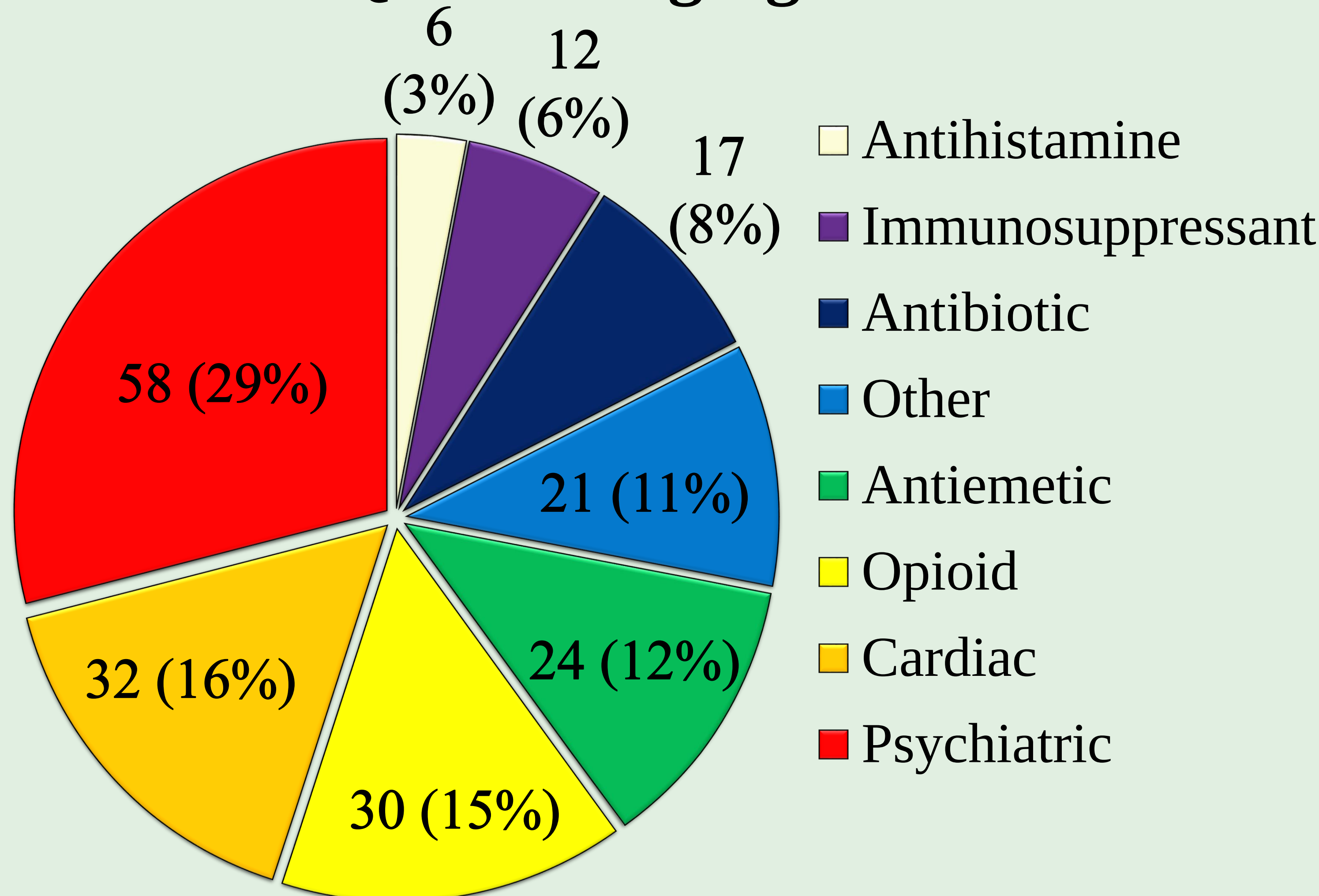
Increased BMI correlated with QTc >440 and QTc>460; increased age, BMI, QRSd, and mortality rate correlated with QTc>460

No patient had ventricular arrhythmia or sudden death during the period of follow-up (6.3 ± 5.1 yrs).

Karyotype	45,X	XX, XO mosaic	Other Karyotype	Unknown Karyotype	All
Number	31	5	30	22	88
BAV	13	2	7	6	28
COA	8	1	0	5	14
AS	3	0	2	0	5
Other Heart DZ	6	1	5	2	14
Any Heart DZ	22	3	14	13	52
Liver DZ	5	4	1	7	17
Hx Cardiac Surgery	12	1	2	6	21
Age (years)	12.97	21.00	17.53	39.64	21.65
BMI	26.92	25.94	32.13	32.17	29.23
Mortality	1	0	1	6	8

Patient	QT-prolonging meds	Non-prolonging meds	No meds	All TS Patients	p value
Number	30	34	24	88	-
Age (avg.)	34.2 $\pm$ 20.8	18.3 $\pm$ 11.0	10.3 $\pm$ 10.6	21.6 $\pm$ 17.8	0.00*
QT (avg.)	370.4 $\pm$ 43.6	377.5 $\pm$ 41.6	340.0 $\pm$ 63.1	365.3 $\pm$ 50.1	0.03*
QTc - Bazette (avg.)	454.8 $\pm$ 31.0	438.7 $\pm$ 16.4	434.4 $\pm$ 33.5	443.0 $\pm$ 28.5	0.01*
QTc - Hodges (avg.)	370.5 $\pm$ 43.5	377.6 $\pm$ 41.6	340.1 $\pm$ 63.1	365.4 $\pm$ 50.5	0.03*
Heart Rate (avg.)	93.6 $\pm$ 21.5	84.1 $\pm$ 11.0	106.0 $\pm$ 10.6	93.1 $\pm$ 17.7	0.01*
QTc B >440	21	15	11	47	0.01*
QTc B > 460	10	4	5	19	0.11

Class of QT-Prolonging Medication



CONCLUSION

Despite the fact that the majority of patients with Turner syndrome have QTc prolongation at baseline, they are commonly prescribed known QT-prolonging medications.

Despite this, no ventricular arrhythmias were identified in this population.

QTc prolongation in Turner syndrome may not carry the same risk of ventricular arrhythmia as seen in patients with long QT syndrome.

REFERENCES

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