A comparison between general rheumatologists and scleroderma experts with respect to following systemic sclerosis guidelines

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ABSTRACT

Objective. To determine if there are differences between expert and non-expert rheumatologists in systemic sclerosis (SSc) management.

Methods. Information relating to demographics, complications, investigations, and treatment of SSc patients was obtained from an online survey to members of the Canadian Rheumatology Association (CRA), and selected chart audits. Results were compared to data from a SSc database ('experts', Canadian Scleroderma Research Group - CSRG).

Results. The online survey (61/300 respondents; 20% response rate) found that most agreed with the EULAR SSc guidelines. Some exceptions were only 47% said they ordered annual echocardiograms and 45% pulmonary function tests. Chart audits of 70 SSc patients from 7 community rheumatology practices revealed no significant differences in their treatment from SSc guidelines, but some investigations differed compared to the CSRG. There was site variability among community practices relating to investigations, and treatment. Patients receiving an echocardiogram within the previous year varied from 10-90%, and PA pressure was reported in 30–100% of SSc patients among sites. Overall, 91% of SSc patients on chart audit had ever received an echocardiogram, but in 30% of cases there was no PA pressure recorded versus only 19% in CSRG (p=0.001).

Conclusion. Compared to SSc experts, general rheumatologists did not differ in their practices for many SSc guidelines despite the fact that they do not see many SSc patients when compared to SSc experts, but there was site variability. An apparent difference is that although echocardiograms are being ordered, PA pressures are missing which could lead to late detection of PAH.

Introduction

Scleroderma (systemic sclerosis; SSc) is a chronic autoimmune connective tissue disease with fibrosis of the skin and other organs, vascular changes and autoantibodies (1). SSc is rare (2), with the majority being middle-aged to older women. SSc is sub-classified into limited (lcSSc) and diffuse cutaneous systemic sclerosis (dcSSc) subsets and both are at risk for internal organ involvement including the GI tract, lung parenchyma and pulmonary arterial hypertension (1, 3-5). It is widely agreed that SSc is one of the most severe forms of connective tissue disease (4); with 10-year survival rates between 80–90% in lcSSc, and 62–76% in dcSSc (5).

SSc is uncommon compared to other rheumatologic diseases. As a result, general rheumatologists who do not have a specific expertise in SSc will see few patients (approximately 14–17 annually) (6). Therefore, it is essential that general rheumatologists have a clear understanding about how to screen for and to treat SSc to improve clinical outcomes. There is evidence supporting treatment of specific manifestations of SSc (7-10) including Raynaud's phenomenon (RP), interstitial lung disease (ILD), and pulmonary arterial hypertension (PAH) (10). The World Health Organization (WHO) made the recommendation of performing annual screening echocardiograms to improve long-term outcomes by detecting SSc associated PAH at earlier, more treatable stages (11). In Canada, there are no protocols or widely accepted SSc guidelines for screening for early SSc organ manifestations (12). Therefore, the majority of tests and treatments that are given to patients are decided at the discretion of the attending rheumatologist on an individual basis. These inconsistencies and variability in managing

SSc have been previously demonstrated (6). However, guidelines relating to the management of scleroderma have recently been published by the European League Against Rheumatism (EULAR) and EULAR Scleroderma Trials and Research (EUSTAR) group based on clinical research evidence and a review of published literature; consisting of 14 recommendations for the treatment of SSc, and addressing specific problems and complications of SSc (13). We have previously studied agreement to the EULAR guidelines by SSc experts in North America and Europe, including members of the Scleroderma Clinical Trials Consortium (SCTC) and the Canadian Scleroderma Research Group (CSRG) (12, 14). Overall, there was strong agreement among SSc experts for many recommendations. Exceptions to expert agreement of the guidelines were the use of iloprost and bosentan for digital vasculopathy, ordering of PAH treatment, and methotrexate use for skin involvement (14). Although expert agreement of the SSc guidelines was generally high, it is unclear whether rheumatologists actually follow these guidelines in practice; as guidelines are usually not followed by significant numbers of specialists. For example, guidelines for the prevention of steroid-induced osteoporosis have adherence between 10–60% of the time (15-17). Our analyses of the practices of SSc experts in the CSRG followed this trend, as many of the EULAR guidelines were only followed 25 –40% of the time (12).

We investigated SSc treatment practices by general rheumatologists in order to determine the agreement with, and variability between the EULAR guidelines and actual practice among general rheumatologists for SSc. The management of SSc was compared between general rheumatologists who performed chart audits and SSc experts from data published from the CSRG database (12)

Methods

This study was made up of three parts. 1. The first was an online survey sent to members of the Canadian Rheumatology Association (CRA). 2. Respondents were invited to perform a chart audit on 10 of their SSc patients and between-site variability was studied. 3. The results were compared to data of treatment and investigation frequencies between the general rheumatologists and larger sites from the Canadian Scleroderma Research group (CSRG) database (12). Site variations from the chart audits were also compared with respect to the frequency of investigations and treatment.

Part 1: An online survey was generated, and sent once to 300 members of the CRA. The questionnaire began with a demographics section (age, sex, year of completion of rheumatology training). (See Appendix A for the questionnaire). It asked the number of patients with SSc that each practicing rheumatologist typically follows, and whether they were aware of the EULAR guidelines relating to the management of SSc (13). We excluded all respondents who did not follow any SSc patients. The remaining questions addressed each participant's current practices relating to diagnosis and surveillance of SSc, and also contained specific questions about the EULAR guidelines concerning treatment of SSc (13). Their perceptions of how they practice were compared to components within the EULAR SSc guidelines (with percentage agreeing to statements).

Part 2: Chart audits were completed for rheumatologists in community practice who responded to the questionnaire and agreed to audit medical records of ten SSc patients whom each participant had seen over the previous 18 months using case report forms for each patient containing questions regarding baseline patient characteristics, SSc-related complications, investigations and treatment.

Part 3: The practice characteristics of the community rheumatolgists who performed the chart audit were compared to published data from the CSRG database (consisting of SSc patients enrolled at several sites in Canada) to determine differences with respect to SSc guidelines. Data that had been previously collected from the CSRG database from 6 centres who had enrolled between 52 and 185 SSc patients were used to compare adherence to guidelines between experts and general rheu-

matologists (18). Comparisons were made using a two-tailed unpaired Student's *t*-test. Differences with *p*-values ≤0.05 were considered statistically significant. These statistical analyses were performed using GraphPad Prism version 3.0 (GraphPad Software Inc, San Diego, CA). Approval was obtained from Western University to perform the research. A comparison of the frequency and variation among rheumatologists who were non-experts in SSc from CRA centres with respect to SSc patients including demographics, investigations and treatment was performed but due to small numbers and multiple comparisons, no statistics comparing sites were performed.

Results

Data from 58 members of the CRA who responded (20% response rate) to the online survey were collected (52% male; with a range of length of time in practice: 44% completed their rheumatology training before 1990 and 21% after 2005). Four other respondents were excluded as they did not follow any SSc patients and they were then told not to complete the survey. Although two-thirds were aware of the EULAR SSc guidelines, only one quarter (26%) had read these guidelines (Table I). The majority of rheumatologists had a small number of SSc patients whom they follow, as only 23% had more than 20 SSc patients. Some agreement (>60% agreement) occurred for the majority of SSc guidelines re screening for internal organ involvement. However, when asked if each respondent ordered annual echocardiograms and PFTs slightly less than half agreed to each. There was strong agreement for most SSc treatment in the EULAR guidelines, with some exceptions such as the ever-use of prostanoids (51%) and bosentan (23%) for digital ulcers. Many respondents thought reminders for ordering echocardiograms, a simple dyspnea questionnaire for SSc patients and a web based form for following SSc may be helpful in their practices (Table I).

Seven rheumatologists from the CRA centres (non-experts) who responded to the survey agreed to perform chart audits; each on 10 patients with SSc

Table I. Responses of CRA to SSc in practice survey.

Survey question	Positive response rate (%) 61/300 (20%)
General	
Are you aware of the EULAR guidelines for management of SSc?	67
Have you read the published EULAR guidelines for management of SSc?	26
Do you follow more than 10 patients with SSc in your practice?	55
Do you follow more than 20 patients with SSc in your practice?	23
Screening and Diagnoses	
Do you screen SSc patients for PAH?	
Always	86
Sometimes	12
Are echocardiograms ordered:	
Annually?	47
At regular intervals but not annually?	25
Only if the patient is symptomatic?	7
Are PFTs ordered:	
Annually	45
Regularly not annually?	33
Only if symptomatic?	7
Do you consistently ask SSc patients about shortness of breath?	76
Are chest x-rays ordered on SSc patients at least once?	81
Do you order HRCT in SSc patients if indicated (suspected ILD, crackles, dyspnea)?	76
Do you routinely ask about GERD?	96
Do you routinely ask about dysphagia?	88
Do you screen for Barrett's esophagitis?	69
Do you routinely ask about Raynaud's phenomenon?	100
Do you ask about and record complications of RP (such as ulcers)?	83
Do you usually do a skin score?	62
Do you record the type of SSc (limited cutaneous *vs.* diffuse cutaneous SSc)?	97
Do you always record BP in SSc patients?	79
Have you ever seen a scleroderma renal crisis?	75
Do you record if tendon friction rubs are present?	68
Treatment	
Are calcium channel blockers used for treatment for SSc-related RP?	100
Do you use prostanoids for treatment of active digital ulcers?	51
Do you consider bosentan for treatment of digital ulcers?	23
Do you use methotrexate in treatment of skin involvement for SSc?	74
Is cyclophosphamide used in the treatment of severe ILD in SSc?	71
Do you use proton pump inhibitors for SSc-related GERD?	69
Unmet need	
Would annual echocardiogram reminders be helpful?	
Yes	35
No	35
Not sure	30
Would it be helpful to have an easy dyspnea questionnaire for your SSc patients?	
Yes	58
No	14
Not sure	28
Would a one-page form on a website help to follow SSc patients?	
Yes	36
Maybe	48
Not sure	16
No	0

EULAR: European League Against Rheumatism; PAH: pulmonary arterial hypertension; PFT: pulmonary function tests; SSc: systemic sclerosis; HRCT: high resolution CT scan of lungs; ILD: interstitial lung disease; RP: Raynaud's phenomenon; GERD: gastroesophageal reflux disease; BP: blood pressure.

whom they were currently following. The data from the chart audits of non-experts were compared to 640 SSc patients within the CSRG database from 6 centres each of whom had entered more than 50 patients with SSc into the CSRG database (experts), with the exception of data from all baseline echocardiograms within the CSRG (n=1319) which were compared to the 7 non-expert centres to determine the frequency of missing PA pressures. Rates of common SSc complications did not differ significantly between CRA and CSRG patients (Table II). Similarly, there were no differences in the treatment practices of physicians between these groups. The frequency of obtaining pulmonary function tests was comparable between groups, although ordering chest x-rays was significantly greater among CSRG-treated patients. Ninety-three percent of all patients in the chart audit had an echocardiogram in the rheumatologists' charts. However, more echocardiograms were done within one year (actually 18 months was used for the CRA members chart audit) but the difference was not significant. PA pressures were lacking in more patients from the chart audit compared to the CSRG database (44% lacking a PA measurement because echocardiogram was not performed or PA pressure was not provided *vs.* 27% in the CSRG database; *p*=0.001). Forty-three percent of patients in the chart audit had an echocardiogram ordered at the current visit. Eighteen percent of the echocardiograms had a PAP of at least 40 mm Hg.

Characteristics for the 7 CRA centres were compared with one another. All of the centres had a majority of female patients and most were lcSSc subset in their SSc chart audit with an average age of between 53 and 66 years (Table III). These data were compared to CSRG patients and when data were unavailable from the key references (18, 12) then other CSRG references were used (25, 26). Other than these baseline characteristics, there was a high degree of site variability among the CRA centres with respect to investigations, SSc-related complications, and treatment. For example, the percentage of patients who received an annual echocardiogram varied from 10 to 90% and the proportion of charts that reported a PA pressure in the echocardiogram report varied from 30 to 100%. Ninety-one percent had ever had an echocardiogram on the chart, but in 1/3 of cases there was no recording of the PA pressure recorded on the echocardiogram and 30% had no echocardiogram in the past 18 months. An echocardiogram was ordered at the current visit in 43% of patients. Eighteen percent had an echocardiogram with PA pressure >40

Table II. Chart audits of scleroderma patients between rheumatologists not expert in SSc (CRA members) and SSc experts (from the Canadian Scleroderma Research Group; CSRG).

Practice comparison	Non-experts in SSc (Chart audits from CRA centres) (n=70) % (frequency)	SSc experts (from CSRG database) (n=640*)[12,18] % (frequency)	*p*-value
SSc Complication			
Raynaud's phenomenon	80	99	0.07
Digital ulcers	60	47	0.4
Dysphagia	61	80	0.8
GERD	71	74	0.8
PAH	6	9	0.3
Treatment			
Calcium channel blockers	44	46	0.7
Promotility agents	19	24	0.5
Proton pump inhibitors	50	64	0.1
Investigation			
CXR	23	90	0.001
Echocardiogram done in last year*	80	95	0.1
PA pressure measured on echocardiogram	56 (39/70)	73 (967/1319)	0.001
(if not echocardiogram was performed, it was assumed not reported)^	(12 no PAP, 19 no echocardiogram)	(227 no PAP, 121 no echocardiogram)	
PA pressure measured on echocardiogram	76 (39/51)	81 (967/1198)	0.4
(including only those that had an echocardiogram performed)^			
PFT done in last year	74	92	0.1

GERD: gastroesophageal reflux disease; PAH: pulmonary arterial hypertension; CXR: chest x-ray; PA: pulmonary artery; PFT: pulmonary function Tests; PAP: pulmonary artery pressure. *CRA members echocardiogram in the last 1 to 2 years, CSRG echocardiogram in the last year, ^PA pressure ever measured in CRA sample and at baseline in CSRG. *CSRG n=640 except for baseline echocardiograms where n=1319 patients.

mm Hg. Approximately 90% had PFTs ever recorded on the chart and 19% had a DLCO % predicted of <60%. Asking the patient about dyspnea was recorded for 64/72 charts (89%). Eighteen percent had a current digital ulcer and more than half of the ulcers were multiple. With respect to complications, very few patients were diagnosed with GAVE (0–10%), Barrett's esophagitis (0–10%), or PAH (with 4 sites having no patients with PAH, two sites 10% of patients and one site 20%).

Discussion

Although Canadian general rheumatologists who completed the survey do not see many SSc patients they reported agreement with many statements from the EULAR SSc guidelines. When comparing EULAR SSc guidelines from actual practice audits of non-experts and CSRG patients, there were many similar practices but there were some differences. There are some apparent practice differences such as missing echocardiograms on the patients' rheumatology charts and/or no measurement of the PA pressure. Only 6% of the total CRA SSc patient population had been diagnosed with PAH. This incidence rate is low with expected PAH in SSc rates between 9% and 12% for PAH proven by right heart catheterisation and 9% in the CSRG database (18, 19). Elevated PA pressures in audited SSc charts occurred in 18% (as echocardiography tends to over or under estimate PAH compared to right heart catheterisation) (20). Interestingly, the percentage of patients who received echocardiograms at CRA centres was fairly high, with nearly all the SSc patients receiving an echocardiogram at least once. Furthermore, these rates did not differ significantly from the CSRG experts (80% *vs*. 95%). This discrepancy suggests that although echocardiograms are being ordered, PA pressure is not measured as often as in the CSRG patients. One study suggested that tricuspid regurgitation was not found on echocardiogram in 9% of cases of PAH (21), but this does not account for the missing PA pressures in 21% (12/56) of echocardiograms. One possible explanation for this discrepancy is that the right heart pressures are not being read and reported. Echocardiograms are currently a screening tool to diagnose PAH in SSc patients, as studies have shown that screening SSc patients with echocardiograms detects PAH at earlier and generally milder stages with an improved prognosis (22, 23). Confirmation with right heart catheterisation is mandatory for the diagnosis of PAH. Therefore, although CRA physicians are ordering echocardiograms for SSc patients, it is essential that they effectively read and interpret them in order to maximise early detection of this serious complication of SSc. The rheumatologists who completed the survey stated that some tools for screening and following SSc patients may be helpful in their practices (echocardiogram reminders, an easy dyspnea questionnaire for SSc patients and an electronic SSc form that could be incorporated into their electronic health records). Measurement of lung function by PFTs was not significantly different in the two groups (but numerically less in the chart audits performed by general rheumatologists) but chest x-rays were not available more frequently in the general rheumatologists *vs*. the CSRG

Table III. A comparison of the frequency and variation among rheumatologists who are non-experts in SSc from CRA centres with respect to SSc patients including demographics, investigations and treatment.

Practice comparison	Experts in SSc (Data from CSRG)[12,18,25,26] n=640 (%)	A (n=10) Frequency (%)	B (n=10) Frequency (%)	C (n=10) Frequency (%)	D (n=10) Frequency (%)	E (n=10) Frequency (%)	F (n=10) Frequency (%)	G (n=10) Frequency (%)
Patient characteristics								
Sex (% female)	87	90	90	60	70	60	70	90
Age (years)	55.3	59.9	52.8	50.6	54.7	65.7	58.1	56.6
% dcSSc subset*	43	10	20	30	50	10	60	40
% lcSSc subset*	57	90	80	60	50	80	40	60
Disease duration (years)	13.7	19.4	8.9	7.0	9.4	8.1	12.4	12.2
Investigation								
CXR ever	72-98	90	10	10	20	10	0	20
HRCT ever	10-41	20	10	0	60	60	100	70
PFT ever	99	90	50	100	100	100	100	90
PFT in last year	92	80	20	80	90	90	90	70
Echocardiogram ever	97-100	100	60	100	100	100	100	80
Echocardiogram last year	95	10	20	90	80	90	90	90
CBC	100	90	70	0	20	70	40	40
Creatinine	100	90	70	0	20	80	40	40
6MWD		10	0	10	0	0	0	0
MD global	100	0	80	0	10	0	0	10
Pain assessed	100	100	20	0	0	0	0	10
Patient global	100	90	20	0	0	0	0	0
ENA	100	90	90	90	100	70	60	80
Upper endoscopy	13 (4-37) ever had a dilation	50	70	0	30	40	90	10
Swallowing study	---	20	10	0	30	0	50	30
PA pressure reported	81	70	50	30	60	90	100	90
RP documented	99-100	50	100	70	100	90	60	100
Asked about dyspnea	100	80	90	60	100	90	100	100
Complications								
GAVE	---	0	0	0	0	10	10	0
Barrett's	---	10	10	0	0	10	10	0
Digital ulcer	58	0	90	10	100	50	100	70
GERD	65-84	30	100	40	100	70	90	70
Dysphagia	84%	40	100	20	100	70	50	50
Skin involvement documented	100 (had MRSS done)	100	100	60	100	70	100	90
PAH	9	0	0	0	20	10	0	10
Treatment								
Calcium channel blockers	60 (ever use) 39-55 (current)	20	50	60	10	60	50	50
Immune suppressives	19 (early dcSSc ever used cyclophosphamide) 25 (8-43) (ever used MTX in early dcSSc) 5-30 (MTX for inflammatory arthritis)	20	30	0	10	30	40	20
Proton pump inhibitors	79-93	50	70	30	70	80	100	40
GI motility drugs	25 (17-33)	10	30	20	10	0	10	50
Steroids	28 (in early dcSSc <3 years) 14=33 (for inflammatory arthritis) 14-26 (if MRSS>10)	10	20	10	30	10	20	0
Treatment changed at current visit	---	20	40	20	10	30	0	30

dcSSc: diffuse cutaneous systemic sclerosis; lcSSc: limited cutaneous systemic sclerosis; *%dcSSc + %lcSSc do not necessarily add up to 100% in each centre as some patients may have been indeterminate or not defined; CXR: chest x-ray; HRCT: high resolution CT scan of lungs; PFT: pulmonary function tests; CBC: complete blood count; 6MWD: 6-minute walk distance; MD: medical doctor (physician); ENA: extractable nuclear antigen; PA: pulmonary artery; RP: Raynaud's phenomenon; GAVE: gastric antral vascular ectasia; GERD: gastroesophageal reflux disease; PAH: pulmonary arterial hypertension; GI: gastrointestinal. --- Data not available; MTX: methotrexate; MRSS: modified Rodnan skin score.
Where data were not available from references 18 and 12, then references 25 and 26 were used to obtain results from the CSRG.

database. The relevance of steroid use is unknown (ranging from 0 to 30% on the chart audits) as we did not collect dose or reasons for prescribing steriods. Treatment with CCBs, PPIs and promotility agents was not significantly different using the EULAR SSc guidelines, but the study may be underpowered to detect differences due to the small number of charts audited. We did study the adherence to practice guidelines using the guidelines and designing a case report form to capture guideline adherence. This is likely a minimum standard of care and cannot ascertain subtle differences in SSc treatment and not all SSc manifestations are covered in the guidelines. We did not request data from right heart catheterisations from the chart audits as we assumed it would not be routinely available on a rheumatologist's chart. Other hints about possible pulmonary hyertension such as falling DLCO over serial PFTs and/or an inappropriately low DLCO such as FVC % predicted/DLCO % predicted ratio of greater than 2 were not captured. We did not ask how a diagnosis of PAH was made. However, specific drugs to treat PAH are not available via provincial access without demonstrating results of the catheterisation.

Data from the CSRG were nearly complete as sites had echocardiogram data entered from 92 to 100% of the time and pulmonary function tests entered 85 to 97% of the time. However there could be tested ordered but not entered into the database (but at low rates due to the nature of data queries within the CSRG) (18). However, data missing from chart audits were truly missing. Thus the CRA members who completed the chart audit were compared to SSc experts with a nearly complete data collection within the CSRG.

Data were collected from the CRA survey in 2010 and the chart audits in 2011; which should have been sufficient time for dissemination of the EULAR SSc guidelines that were published in 2009 (13).

The survey had a poor response rate. The CRA organisation agreed to send it to their members only once. There is no CRA members' directory and obtaining email addresses of Canadian members

of the American College of Rheumatology would potentially favour dissemination of academic rheumatologists. The response rate is in the range of what would be expected for a survey to members of an organisation asking about a rare disease. Involvement in the online questionnaire and the chart audit for CRA physicians was voluntary. CRA members are both community and academic. We did not collect data on practice type. Respondents had approximately equal numbers in those who completed rheumatology training from 1980 to 2005 (10–15% in each category) and 21% completed training after 2005 (so a few more junior members were represented). After excluding 4 who follow no SSc patients, we found that 40% of respondents follow less than 10 SSc patients annually, 1/3 between 10 and 20 patients, and only 1/5 followed 21 or more patients with SSc annually so the majority were not SSc experts if expertise can be measured by the number of SSc patients followed in a year. The generalisability to the CRA members as a whole is that younger rheumatologists were slightly over-represented on our survey, but the usual number of SSc patients followed currently by our CRA members is not known. However, in past we published that the usual CRA member sees approximately 14 patients with SSc annually (6). Therefore, our findings likely highlight a best case scenario with regards to SSc investigations and treatment than would be seen in usual rheumatologic care (as physicians were self-selecting). Chart audits and responses to the survey questions were not verified by an independent observer. Physicians may report they are doing better on a survey than what an actual audit of their practices may reveal as most of the respondents did not choose to perform an audit. In chart audits, the time frame of the last 18 months may have been extended when completing the forms. Practices may be different now as the standard of care in SSc is changing and this could be reassessed in the future. Guidelines will likely be updated to reflect these changes.

Educational studies have suggested that physicians may benefit from regular

and routine feedback in order to assist them with self-assessment and improvement (24). A chart audit allows a professional to reflect on their practice but without a random sample it is uncertain whether the results of the audits are generalisable to other rheumatologists. This study addresses comparisons of a small number of general rheumatologists who on the survey did not verify their answers by a chart audit (response and reporting biases) and a small group who performed a chart audit who may not be representative of usual rheumatologists. The extraordinary complex care SSc patients require for the spectrum of varied manifestations could not be ascertained.

Conclusions

General rheumatologists do not follow as many patients with SSc but they have practices that are similar to experts in SSc care for several recommendations from the EULAR SSc guidelines. There may be differences where screening echocardiograms do not measure the PA pressure at non-expert centres. Perhaps when ordering screening echocardiograms on SSc patients it should be stressed that 'estimating the PA pressure and ruling out PAH' could help to ensure that PAH is screened for and PA pressures reported more routinely. There were also wide ranges of the frequency of some investigations and treatment when comparing the practices of non-experts. Some tools may aid in the care of SSc for the general rheumatologist such as case report forms that can be entered into electronic medical records, or reminders such as when to perform regular screening tests such as echocardiograms and pulmonary function and/or prefilled echocardiogram forms to have measurements performed for estimating PA pressure may be helpful but were not studied in the chart audits.

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