

All patients had plain X-ray skull including craniovertebral junction and CT scan of head to rule out craniovertebral anomaly, tumour or hydrocephalus. Symptoms suggestive of autonomic dysfunction were enquired into. All subjects were screened to exclude diabetes mellitus, adrenal and thyroid disorder, chronic alcoholism and prolonged drug intake, factors likely to alter the results of autonomic nervous function tests.

An informed consent was obtained from the patients after explaining the procedure in detail. The subject adopted a comfortable posture. ECG electrodes were placed and connected to the Biorite (INCO, Ambala, India). The other channel of Biorite was used to record respiration through stethograph. The ECG (lead II) and respiration were recorded throughout tests. The following non-invasive tests were carried out (13).

1. *Isometric exercise* : The subject pressed the hand grip dynamometer for four min at 30% of the maximal voluntary effort. Adequate effort by all patients was ensured. Blood pressure (BP) was recorded serially (at 1, 2, 4, 6 min) in the contralateral arm. Heart rate was monitored by ECG.

2. *The cold pressor test* : For this test, both feet of the patient were immersed in cold water at 10°C for 1 min (14). The BP and HR were recorded simultaneously at 1, 2, 5 and 10 min after immersion.

3. *Head-up tilt test* : After recording the baseline BP and HR while supine on the tilt table, the subject (duly strapped) was passively tilted head up to 70° in 15 sec and serial recordings of BP at 0.5 min, 1 min, 2.5 min, 5 min (tilted), and 5.5 min (horizontal), 7.5 min and 10 min were noted.

4. *Heart rate variation with respiration* : Patients were instructed to regulate their respiration to 6 breaths per min in the sitting position. The ratio of the 6 longest to the 6 shortest R-R intervals was determined

(E:I ratio). The difference between the maximum and minimum heart rates was also calculated.

5. *The valsalva maneuver* : The seated subject was asked to expire for 15 sec to raise and maintain a pressure of 40 mm Hg in a closed system. The Valsalva ratio (VR) was calculated as the ratio of the maximum R-R interval during phase IV to the minimum R-R interval during the phase II.

Various indirect autonomic parameters obtained from patients and controls were compared by Student's unpaired 't' test. P values were calculated and considered significant when $P < 0.05$.

RESULTS

The mean age of ADCA patients was 39 ± 11 years. Only three patients had symptoms suggestive of autonomic dysfunction. These three patients had history of constipation, two of them reported impotence and one of them had hesitancy at micturition and precipitancy of urine. Table I and II present the raw data of autonomic responses. The controls were 20 normal subjects with no history of alcoholism (mean age 35 ± 8). The control subjects did not have any of the autonomic symptoms.

Comparison of patients and controls for autonomic responses revealed that the two had no significant differences for heart rate response for five autonomic tests that we used. The pressor response showed significant differences. The diastolic pressure showed significantly less rise during hand grip test and cold pressor test ($P < .001$). A significant fall in diastolic blood pressure was noticed during head-up tilt ($P < .001$). A significant less rise in systolic blood pressure in both the tests (hand grip test and cold pressor test) was seen although at lower level of significance ($P < .01$). Change in systolic pressure was not significantly different in two groups during head-up tilt. The values for E:I ratio, Valsalva ratio and 30:15 ratio were not different for controls and patients (Table I, II).

TABLE 1 : Autonomic responses for ADCA patients.

S.No.	Age (yrs)	E:I ratio	Vals. ratio	<i>Hand-grip test</i>				<i>Cold pressor test</i>				<i>Head-up tilt</i>				
				<i>Change in blood pressure</i>				<i>Change in blood pressure</i>				<i>Change in blood pressure</i>				
				SBP	DBP	%SBP	%DBP	SBP	DBP	%SBP	%DBP	SBP	DBP	%SBP	%DBP	Ratio
1.	36	1.28	1.70	30	18	25.90	25.70	26	24	23.60	34.30	-10	-08	-8.77	-11.10	1.08
2.	38	2.33	1.85	04	06	03.07	06.66	02	04	01.53	04.44	-10	-06	-7.93	-07.50	1.11
3.	44	1.23	1.16	12	14	11.50	20.00	00	-04	00.00	-05.71	-04	-10	-3.84	-14.30	1.05
4.	41	1.18	1.33	24	10	13.30	10.00	00	04	00.00	00.40	-06	-04	-3.40	-04.25	1.03
5.	38	1.08	1.40	20	10	15.40	11.10	10	06	07.69	06.66	-10	-10	-7.70	-11.10	1.03
6.	36	1.28	1.70	16	14	11.80	15.90	-04	08	-02.90	09.10	-02	-08	1.50	-09.10	1.04
7.	32	1.19	1.47	10	10	20.00	12.50	00	06	00.00	07.50	-10	-10	-10.0	-12.50	1.04
8.	38	1.19	1.91	04	02	03.44	02.32	12	14	10.30	16.30	-10	-06	-7.70	-16.97	1.08
9.	50	1.20	1.18	24	18	19.40	24.30	16	06	12.30	06.97	-12	-06	-8.80	-07.30	1.05
10.	28	1.17	1.55	06	-04	04.16	-04.25	-18	-04	-12.50	-04.30	-10	-00	-7.10	00.00	1.23
11.	39	1.44	1.86	-04	-04	02.85	-04.25	04	00	02.85	00.00	00	00	0.00	00.00	1.03
12.	38	2.00	2.30	30	14	26.80	17.90	-02	02	-01.66	02.43	-28	-12	-15.5	-16.20	1.03
13.	46	1.18	1.53	16	02	10.70	01.92	10	06	06.66	05.76	-10	-04	-6.70	-04.00	1.04
Mean ± SD	39 ±11	1.61 ±0.16	1.91 ±0.33	14.76 ±10.60	8.46 ±7.53	22.74 ±08.34	28.05 ±11.70	4.3 ±10.7	5.53 ±7.30	17.71 ±7.18	20.20 ±8.19	-9.38 ±6.70	-6.46 ±3.75	5.24 ±6.89	5.82 ±10.80	1.07 ±0.03

E:I ratio, expiration-inspiration ratio; Vals., Valsalva; SBP, Systolic blood pressure; DBP, diastolic blood pressure.

For hand-grip test given values are at 4th min, for cold pressor test at 1st min and for head-up tilt the values are maximum changes during five min of tilt.

TABLE II : Autonomic responses for normal subjects.

S.No.	Age (yrs)	E:I ratio	Vals. ratio	<i>Hand-grip test</i>				<i>Cold pressor test</i>				<i>Head-up tilt</i>				ratio
				<i>Change in blood pressure</i>				<i>Change in blood pressure</i>				<i>Change in blood pressure</i>				
				SBP	DBP	%SBP	%DBP	SBP	DBP	%SBP	%DBP	SBP	DBP	%SBP	%DBP	
1.	26	1.67	2.39	18	26	14.28	34.21	20	14	15.87	18.42	-04	-10	-3.17	-13.15	1.06
2.	26	1.55	1.67	10	06	10.00	08.33	16	16	16.00	22.22	00	-02	0.00	-02.77	1.03
3.	28	1.60	2.63	44	34	41.50	51.51	16	14	15.09	21.21	-10	00	-9.43	00.00	1.09
4.	28	1.54	1.55	32	26	28.57	35.13	20	08	17.85	10.81	-06	-02	-5.35	-02.70	1.06
5.	29	1.59	1.52	12	12	12.00	17.14	14	10	14.00	14.28	-04	06	-4.00	08.57	1.09
6.	29	1.23	1.76	26	26	26.53	44.82	18	12	18.36	20.68	-08	-14	-8.16	24.13	1.12
7.	30	1.50	2.20	22	12	19.29	16.66	18	14	15.78	19.44	02	08	1.75	11.10	1.09
8.	31	1.67	1.54	30	40	26.78	50.00	28	24	25.00	30.00	-20	12	-17.85	15.00	1.15
9.	32	1.66	2.00	18	22	16.36	31.42	08	18	07.20	25.71	10	10	09.09	14.28	1.05
10.	34	1.67	2.08	24	24	21.81	31.57	14	14	12.72	18.42	-10	16	-09.09	21.05	1.09
11.	38	1.60	2.41	18	16	15.00	17.77	-04	-02	-03.33	-02.22	-02	06	-01.66	06.66	1.07
12.	46	1.58	2.03	30	22	25.86	28.20	24	16	20.68	20.51	-22	10	-18.96	12.82	1.03
13.	46	1.50	1.58	10	10	07.93	12.50	32	20	25.39	25.00	-10	-06	-07.93	-07.50	1.03
14.	45	1.36	1.67	08	16	06.68	20.00	26	16	21.66	19.51	-06	02	-05.08	02.56	1.09
15.	49	1.56	1.75	40	28	30.76	29.96	00	10	00.00	10.41	00	10	00.00	12.50	1.10
16.	40	1.45	1.96	20	10	15.62	10.63	24	24	18.46	25.53	08	04	06.55	04.16	1.06
Mean	35	1.54	1.92	22.62	20.62	19.93	27.49	17.12	14.25	15.04	18.74	-5.12	3.12	-4.58	6.66	1.07
± SD	±7.9	±0.12	±0.34	±10.60	±9.4	±9.05	±3.60	±9.57	±6.27	±7.94	±7.65	±8.57	±8.32	±7.55	±10.20	±0.03

(Abbreviation : Same as for Table I)

DISCUSSION

Three of our 13 patients of ADCA had symptoms of autonomic dysfunction. All 3 had constipation, and one of them had bladder dysfunction in the form of hesitancy and precipitancy. Kuroiwa (15) found five out of eleven patients of OPCA with postural hypotension and Miyazaki (16) documented orthostatic hypotension in 20 patients out of 45 patients with cerebellar degeneration. Chokroverty et al (17) reported autonomic deficits in 4 out of 10 patients of olivopontocerebellar degeneration.

The patients did not differ from normals for parasympathetic reactivity (as indicated by E : I ratio, Valsalva ratio and 30:15 ratio). This indicates towards normal parasympathetic functions in ADCA patients. The pressor responses in ADCA patients were definitely lower than controls (Table II). The diastolic pressure during hand grip and cold pressor tests were significantly lower than normals suggesting a decreased sympathetic drive during test situation. This fact is further confirmed by significant fall in diastolic blood pressure during head up tilt. The decreased sympathetic drive should be accompanied by reduced rise in systolic

blood pressure during the hand grip and cold pressor test. This is what was precisely observed for systolic pressure (Table II). Thus, these patients have a tendency towards postural hypotension albiet none had clinical manifestations.

Major studies on ADCA or OPCA have neglected the study of the autonomic functions even though the involvement of pons, medulla, neurotransmitter, and receptor systems is well known. The autonomic fibres could well be damaged in the brainstem and subclinical autonomic neuropathy may be a part of ADCA. In our patients of ADCA, autonomic function testing indicates involvement of the sympathetic pathways. Knowledge of the autonomic dysfunction in ADCA cases may have therapeutic and prognostic implications. It may be interesting to investigate different types of cerebellar ataxias for autonomic dysfunction as all these patients undergo extensive physiotherapy and rehabilitative measures.

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