Title: Interval CT coronary angiography in 15children with Kawasaki disease:Our experience at Chandigarh, North India

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Background: Coronary artery abnormalities (CCAs) can occur in ≈25% of children with KD. Transthoracic 2D-Echocardiography (2DE), hitherto the imaging modality of choice, has several limitations as an imaging modality. Computed Tomography Coronary angiography (CTCA) has enabled the comprehensive evaluation of coronary arteries in children with KD. This study pertains to intervalCT coronary angiography in 15children with KDat a tertiary care centre in Chandigarh, India

Patients and methods:CTCA was carried out on 128-slice dual-source CT scanner(Siemens, Erlangen, Germany). There were 15 children in whom interval CTCA was performed.

Results: Median age at diagnosis of KD was 48 months [range 4-96 months]. Median interval between the first and second CTCA examination was37 months [range 6-85 months]). Findings of CTCA at presentation revealed21 aneurysms and 11 dilatation: left main coronary artery (LCA) –5 aneurysm and 2 dilatation and ectasia in 1;left anterior descending artery (LAD) – 9 aneurysm, 1 dilatation;right coronary artery (RCA)–7 aneurysm and 1 dilatation andleft circumflex artery (LCx) dilatation in 6 patients. Giant aneurysms were present in6patients (LAD – 5; RCA-3). Thrombosis and stenosis were noted in 3patients each in initial CTCA and complex aneurysm with multiple skip lesions are seen in 3 patients. Interval CTCA was completely normalized in 5/15 (33.3%) patients. Remaining 10 patients showed persistent (albeit regressed/remodelled) coronary artery aneurysms: LCA-5; LAD-8; RCA-7; LCx-2.Two patients had shown mural calcifications. Two patientsin whom CTCA was performed at intervals

of37months and 72 months after diagnosis of KD, revealed long segment stenosis in LAD and significant mural calcification. One patient developed thrombus in fusiform aneurysm of LAD after 42 months. Of the 3 patients with stenosis in initial CTCA, 2 patients had normal study after 60 months of follow up, one patient had progressive stenosis involving up to 80% of LAD. One patient developed stenosis in follow up which wasn't noticed in the initial CTCA.

Conclusions: Children with KD and CAAs require prospectivelong-termfollow-up as they may develop complications like thrombosis, stenosis, and calcifications.CTCAprovides more detailed and comprehensive evaluation in comparison to 2DE.

Incomplete Kawasaki disease with Giant coronary aneurysms: The usefulness of BCG reactivation as a diagnostic tool

Abstract:

Introduction : Kawasaki disease (KD) a medium vessel vasculitis of unknown etiology.It is an acute febrile multisystem disorder. Most commonly affected children are younger than 5 years of age and it is less common condition in infants younger than 3 months old with paucity of signs/symptoms.. Unfortunately, cardiovascular complications are most common in young infants and incomplete KD is most frequently reported in this age group.

Methods : We report a 4 months old female infant with history of fever , irritability , cracking of lips and edema over hands and feet.There wasinflammatory reactivation of Bacillus Calmette Guerin (BCG) scar in left upper arm in deltoid region (Figure).

Results : Laboratory investigations done revealed raised inflammatory markers (ESR - 110 mm/hr, CRP- positive and platelets - 437,000/mm^3). On the basis of history, clinical signs and laboratory findings, a possibility of KD was considered and she received IVIG @ 2gm per kg and medium dose aspirin was started.2D echo revealed LMCA and left anterior descendingArtery (LAD) dilatation of 4mm each with Z score of 6.41 and 12.81 respectively. In view of giant aneurysm child received infliximab and was started on low dose aspirin and anticoagulant therapy.

Discussion : As there is a dilemma in diagnosing incomplete KD in infancy, any sign helping in early diagnosis of KD , such as BCG reactivation is useful.

Conclusion :BCG reactivation helped us in making the diagnosis of KD, with a highlight on usefulness of this sign for early diagnosing incomplete KD, especially in countries where BCG vaccination is part of immunization schedule.

Discrepancies in identifying and quantifying coronary dilatation among standard z score nomograms in children with Kawasaki Disease, and Multisystem inflammatory syndrome-A Descriptive Study

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ABSTRACT

Background: Kawasaki disease (KD) is an acute vasculitis that preferentially affects medium sized arteries, particularly the coronary arteries (CA).CA dilatation wasalso observed in a proportion of children who presented with KD like manifestations during the COVID-19 pandemic, a condition later named asPediatric Inflammatory Multisystem syndrome temporally associated with SARS-CoV-2 (PIMS-TS) or Multisystem inflammatory syndrome in children (MIS-C) (1).CA dilatation in KD has been graded based on the amount of dilatation for therapeutic decision making regarding the need for treatment with intravenous immunoglobulin (IVIg) and long-term management with regard to need for additional therapy with anticoagulants. The initial Japanese Health Ministry sizing scheme (2)which was based on absolute CA dimension according to dichotomous criteria of age (</>= 5 years), was later replaced by CA size grading by z scores based on body surface area (3,4,5). Standard works of reference done on normal children have provided-echocardiographic z score nomograms for coronary artery dimensions in children that can be used to derive z scores of patients (6-10), however there is no consensus as to which standard is appropriate for Indian children. Since therapeutic decisions at many stages in the treatment of KD are based on z score of coronary artery sizes, it is important that we use a reliable, standard nomogram that has been validated and ascertained to be appropriate for our population.

Although diagnosis is based on clinical criteria, the decision to treat with IVIg, especially when treating incomplete KD is based on Z scores of left anterior descending (LAD) coronary artery or Right coronary artery (RCA) dimensions (4). In children with large coronary aneurysms, the need for starting anticoagulation therapy is decided by z score > 10 (4). Since the guidelines do not recommend any specific nomogram, the Z score method has to be decided by the treating physician. Our study tried to compare the CA dimensions in normal as well as KD/MISC children based on the Z scores calculated by five standard methods: Boston, Washington, Montreal, Kobayashi and Pediatric Heart Network (PHN) (6-10).The Coronary Aorta index (CAI) (8,11) which is the ratio of coronary artery to aortic annulus, was also assessed for every subject and compared with the subject's coronary artery z scores.

Aims and objectives : The primary objective is to determine the agreement between Z scores derived from the existing nomograms for CA dimensions including that derived from coronary aorta index (CAI) in children belonging to the age groupof 1 month - 10 yearswho are/were diagnosed with KDin our Institute. The secondary objective is to determine the agreement between Z scores derived from the existing nomograms for CA dimensions including that derived from coronary aorta index (CAI) in children belonging to the age groupof 1 month - 10 years who are/were diagnosed with MIS-Cin our Institute and compare it with normal controls.

Patients and methods : This descriptive study included three broad groups of normal, KD, and MISC children aged 1 month to 10 years. Normal children were recruited from immunization OPD and from pediatrics OPD/IPD who presented with some other ailments.Children with congenital or acquired cardiomyopathy, congenital heart disease, other vasculitis includingsystemic lupus erythematosus, immunosuppression, severe sepsis and multi organ dysfunction syndrome were excluded from this study. ECG gated echocardiographic examinations were performed in the Department of Cardiology using Philips iE 33(Philips Healthcare, Andover, MA, USA) or Philips EPIQ 7 (Philips Healthcare, Andover, MA, USA) ultrasound Machine using S 12- 4 or S8-3 probes. The proximal right coronary artery (RCA), left main (LM), left anterior descending (LAD) and left circumflex (LCx) coronary arteries were measured in the parasternal short axis view

in a mid diastolic frame. LM was measured midway between its ostium and its bifurcation into LAD and LCx. LAD, LCx and RCA were measured 3-5 mm distal to their origin (12). The measurements were assigned z scoresusing 5 standard nomograms which are routinely being used for this purpose. For LCx dimensions, z scores are provided only by Dallaire et al (8) and Kobayashi et al (9). Hence only these 2 z scores were assigned for the measured LCx dimension. For LM, LAD and RCA dimensions, all nomograms were employed and the differences were assessed. Aortic annulus was also measured and the Coronary aorta index (CAI) (8,11) was calculated and the respective z score were computed using the reference equations described by Dallaire et al (8).

Statistical analysis: Diagnostic accuracy of each z score nomogram in comparison to the results from Boston z score nomogram was assessed using sensitivity, specificity and area under the curve.Analysis of variance (ANOVA) was used to compare coronary artery Z scores between the three groups. The agreement between CA measurement z scores and z scores of coronary aorta index) was assessed using Bland-Altman plots. All the statistical analysis were carried out at 5 % level of significance and a p value < 0.05 was considered significant. The sample size was estimated based on the estimation of sensitivity of each of the z score nomograms in diagnosing the children with and without Kawasaki disease from heathy children. The recruitment of Kawasaki children was taken in the ratio of 1:4 with respect to healthy children. Therefore, at 5% level of significance, with an anticipated minimum sensitivity of the nomogram to be at 90 %, and a margin of error of 0.15, a total of 56 Kawasaki patients was estimated to be sampled after adjusting for multiple comparison using Bonferroni correction. Equal number of MISC patients in the ratio of 1:1 to Kawasaki patients were selected in the study. Hence, the number of healthy children in the study was estimated to be 223 and therefore the total sample size for the study was estimated to be 335.

Results: Total 275 children were included in this study till now, 69.1% in normal group, 13.5% and 17.5% in the KD and MISC groups respectively. The mean age of the children was 62.58 months and 59.6% were males. The frequency distribution of

categorical and continuous variables are shown in Table 1 and 2 respectively. The agreement between the Z scores derived by Boston method and those derived from other methods including the coronary aorta index was analysed and shown in Tables 3-42 and Figures 1-42. Amongst normal children, the agreement between LMB and LMW were assessed using intraclass correlation (ICC). It was found that the ICC r value was 0.983 (95% confidence interval: 0.978, 0.987) which was found to be statistically significant (P value < 0.001). This suggests a very high agreement between the methods. A Bland Altman (BA) plot was plotted to visually represent the agreement between the methods. It was found that 12 (6.35%) of children were outside the limit of agreement between these two methods. Similarly the agreement between z scores by various methods was also assessed for KD and MISC group. The best agreement was between Boston and PHN methods for LM and LAD and between Boston and Montreal methods for RCA in the normal group with the ICC r values of 0.993, 0.999 and 0.985 respectively. In both KD and MISC groups the best agreement was between Boston and PHN methods for LAD and between Boston and Montreal for RCA. For LM in KD group agreement was better between Boston and Montreal methods with the ICC r value of 0.998 and between Boston and Washington metgods in the MISC group with ICC r value of 0.992.For LCx agreement was seen between the Z scores derived from Montreal regression equations with the square root of body surface area and those derived from equations using aortic annulus diameter. This assessment suggested a comparatively lower agreement in the normal group (ICC r=0.882) compared to that in the KD and MISC group (ICC r=0.987, 0.909). For RCA, LAD and LM also similar results were seen with the highest agreement in the KD group for the 2 methods.

Conclusions: In our study we found that various coronary artery Z score formulas have high agreement with Boston Z scores in all three groups of normal, KD and MISC children. On comparing Z scores derived from aortic annulus diameter equations to that derived from Montreal regression equations with square root of body surface area highest agreement was seen in the KD group but the agreement was relatively lower in the normal children for LAD, RCA and LCx and in MISC group for LM.

Table 1. **Frequency distribution of categorical variables**

A. GROUP

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Normal	190	69.1	69.1	69.1
	KD	37	13.5	13.5	82.5
	Misc	48	17.5	17.5	100.0
	Total	275	100.0	100.0	

B. SEX

		Frequency	Percent	Valid Percent	Cumulative Percent
Valid	Female	111	40.4	40.4	40.4
	Male	164	59.6	59.6	100.0
	Total	275	100.0	100.0	

Table 2. Frequency of continuous variables

Case Summaries

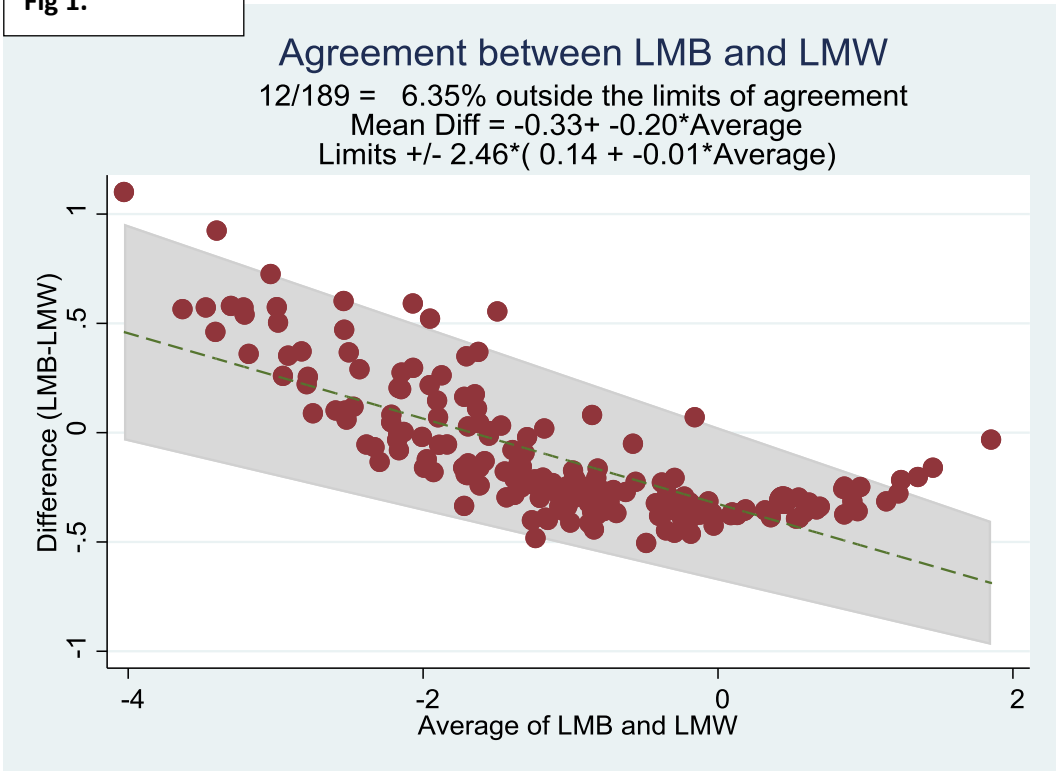
	AGE	HT(cm)	WT(kg)	Ao Ann	LM	LAD	LCX	RCA
N	275	275	275	257	274	275	274	275
Mean	62.587	105.219	18.14807	12.6616	2.1841	1.5072	1.2623	1.4745
Std. Deviation	41.9397	26.3999	10.288413	2.83810	.72041	.51846	.45307	.63895

In normal children, agreement of LMB with LMW

Table 3. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.967[b]	.956	.975	59.539	188	188	.000
Average Measures	.983[d]	.978	.987	59.539	188	188	.000

Fig 1.

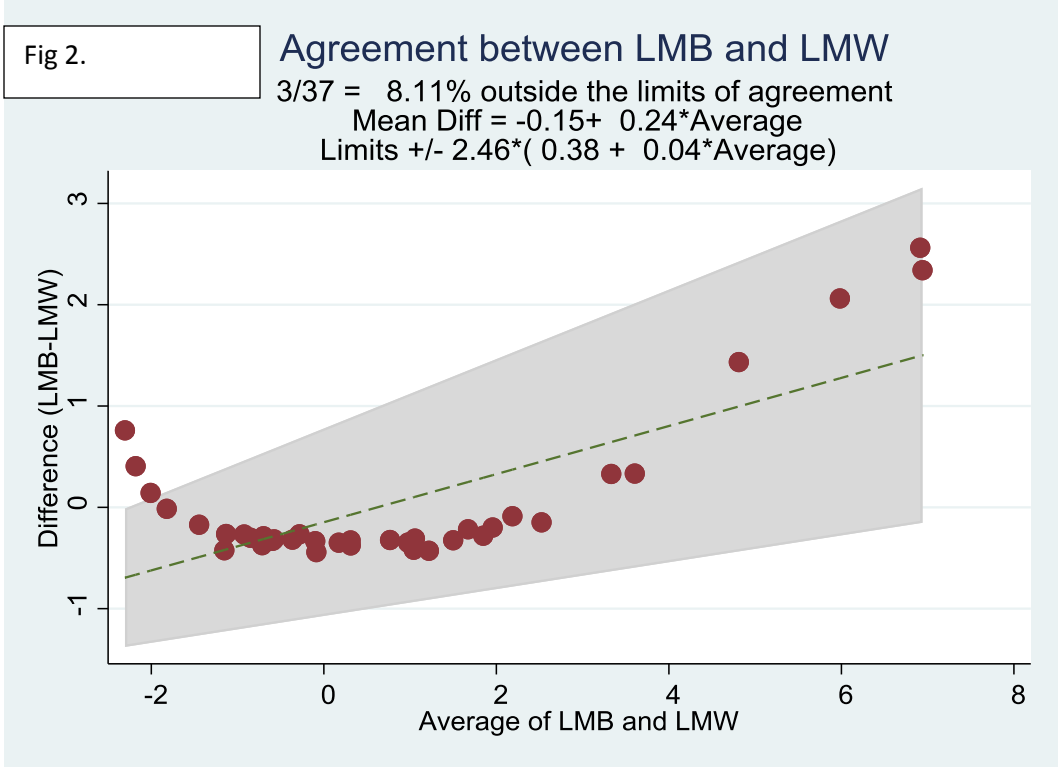


In KD children, agreement of LMB with LMW

Table 4. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.948[b]	.902	.973	37.608	36	36	.000
Average Measures	.973[d]	.948	.986	37.608	36	36	.000

Fig 2.

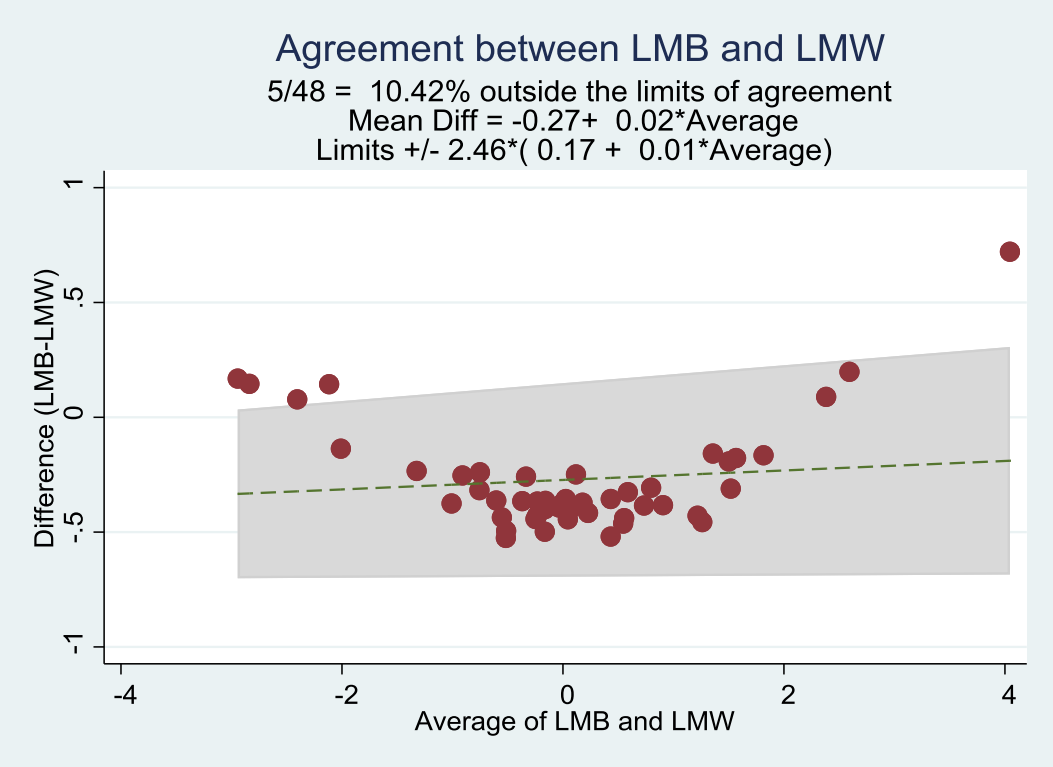


In Misc children, agreement of LMB with LMW

Table 5. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.984[b]	.972	.991	125.208	47	47	.000
Average Measures	.992[d]	.986	.996	125.208	47	47	.000

Fig 3.

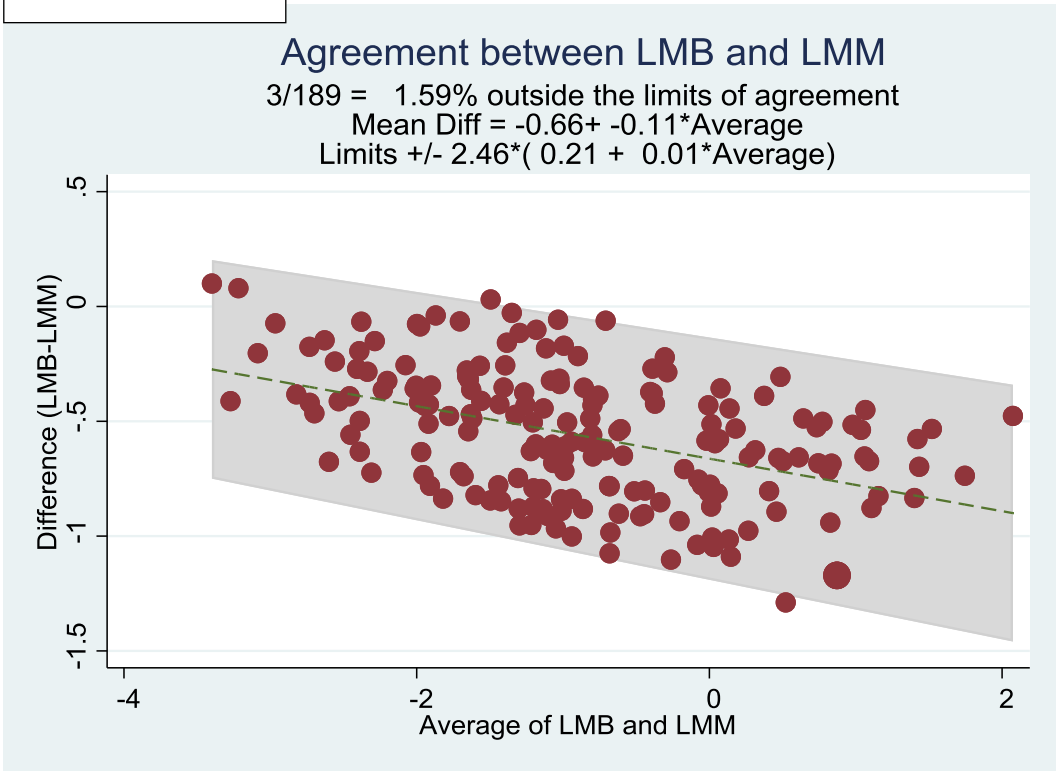


In Normal children, agreement of LMB with LMM

Table 5. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.969[b]	.959	.977	63.998	188	188	.000
Average Measures	.984[d]	.979	.988	63.998	188	188	.000

Fig 4

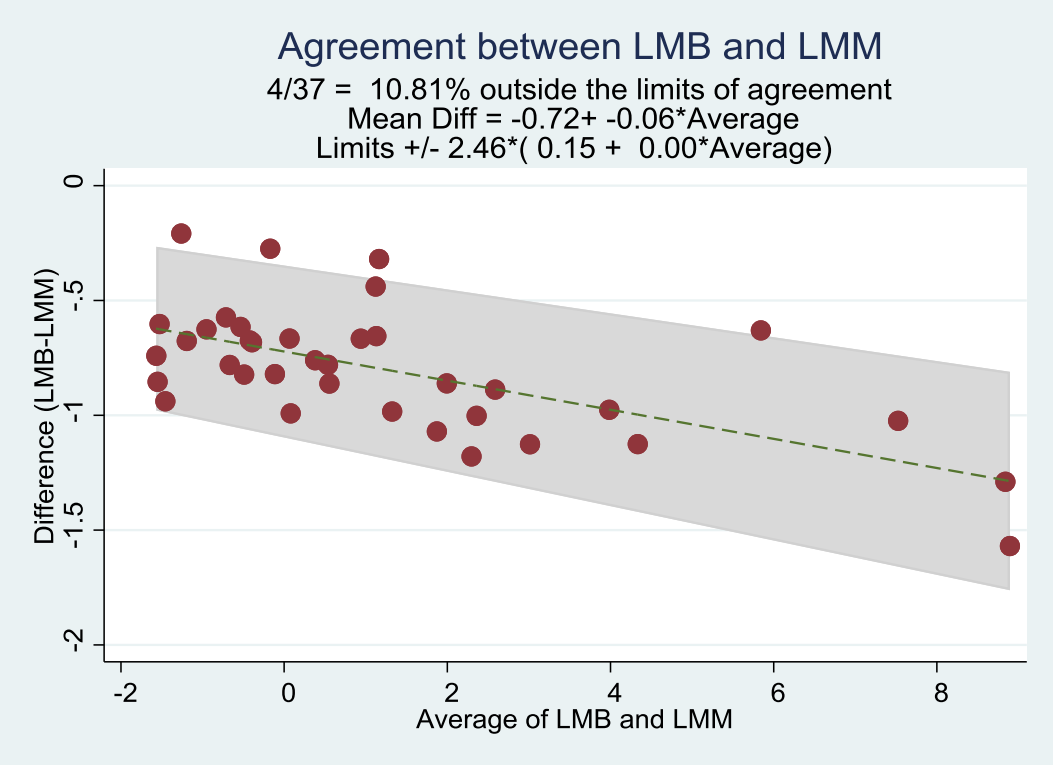


In KD children, agreement of LMB with LMM

Table 6. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.995[b]	.990	.997	400.558	36	36	.000
Average Measures	.998[d]	.995	.999	400.558	36	36	.000

Fig 5

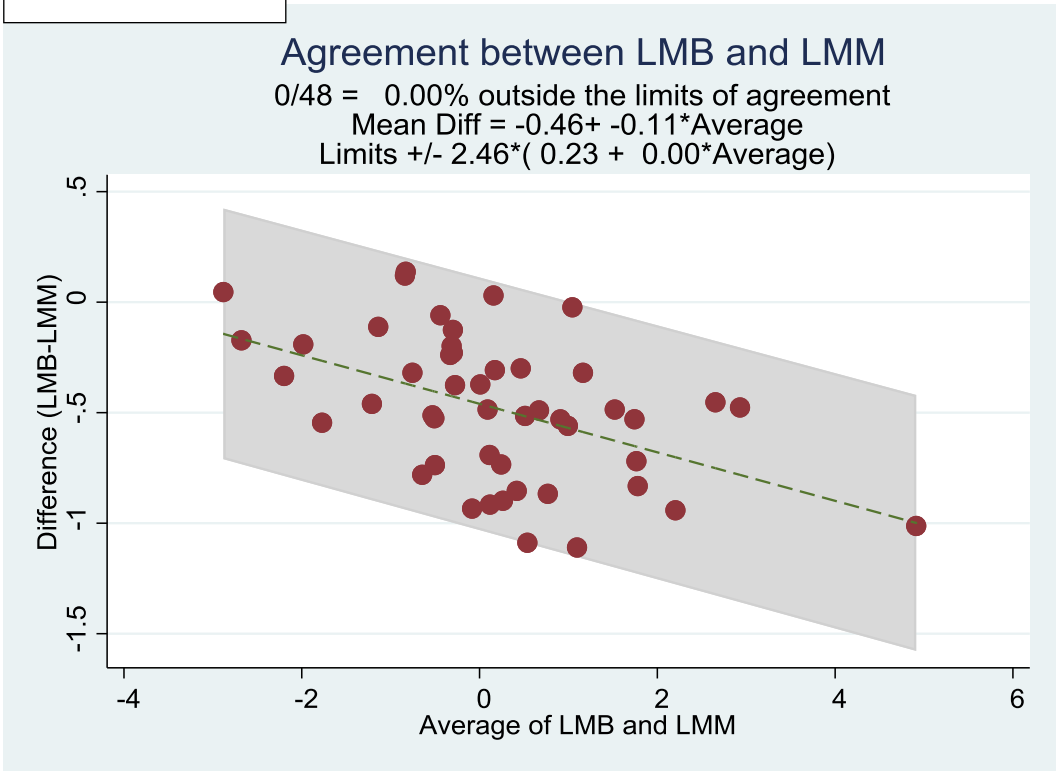


In Misc children, agreement of LMB with LMM

Table 7. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.974[b]	.954	.985	76.188	47	47	.000
Average Measures	.987[d]	.977	.993	76.188	47	47	.000

Fig 6

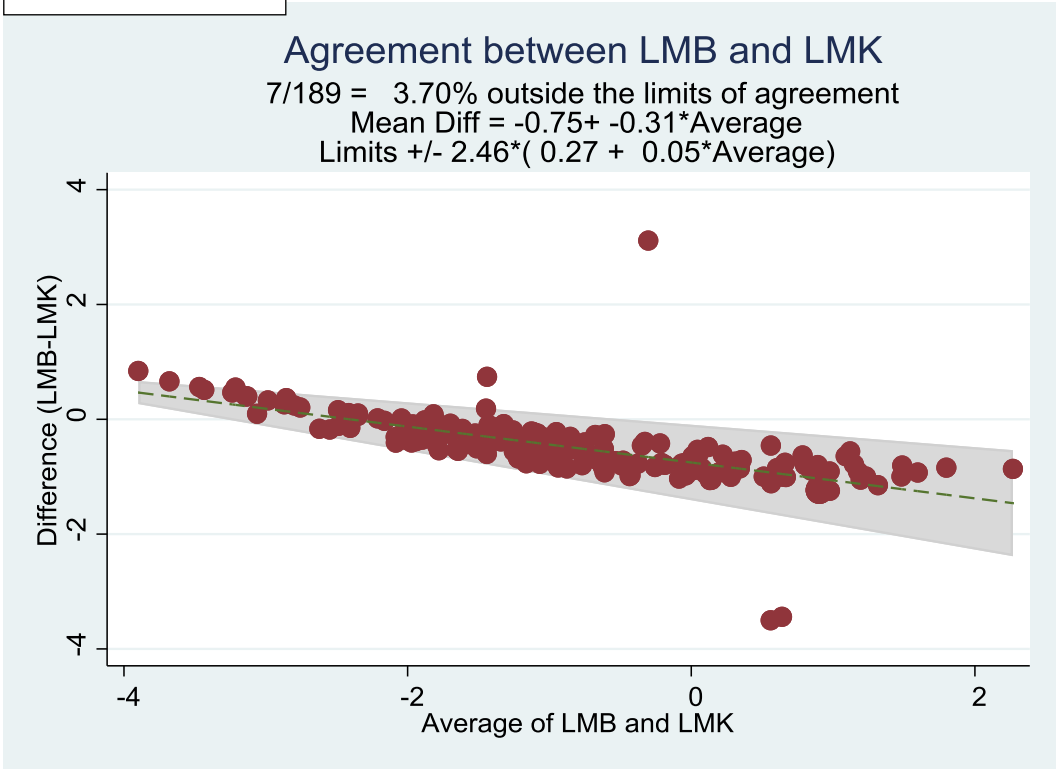


In normal children, agreement of LMB with LMK

Table 8. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.894[b]	.862	.920	17.948	188	188	.000
Average Measures	.944[d]	.926	.958	17.948	188	188	.000

Fig 7

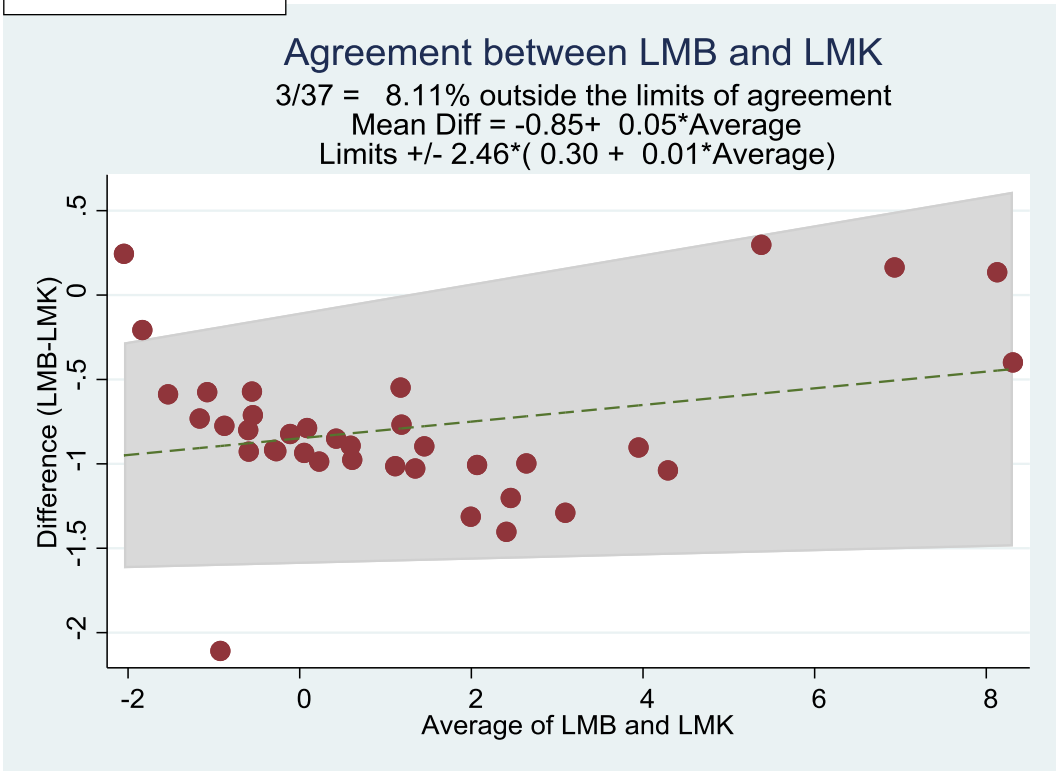


In KD children, agreement of LMB with LMK

Table 9. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.984[b]	.969	.992	123.661	36	36	.000
Average Measures	.992[d]	.984	.996	123.661	36	36	.000

Fig 8

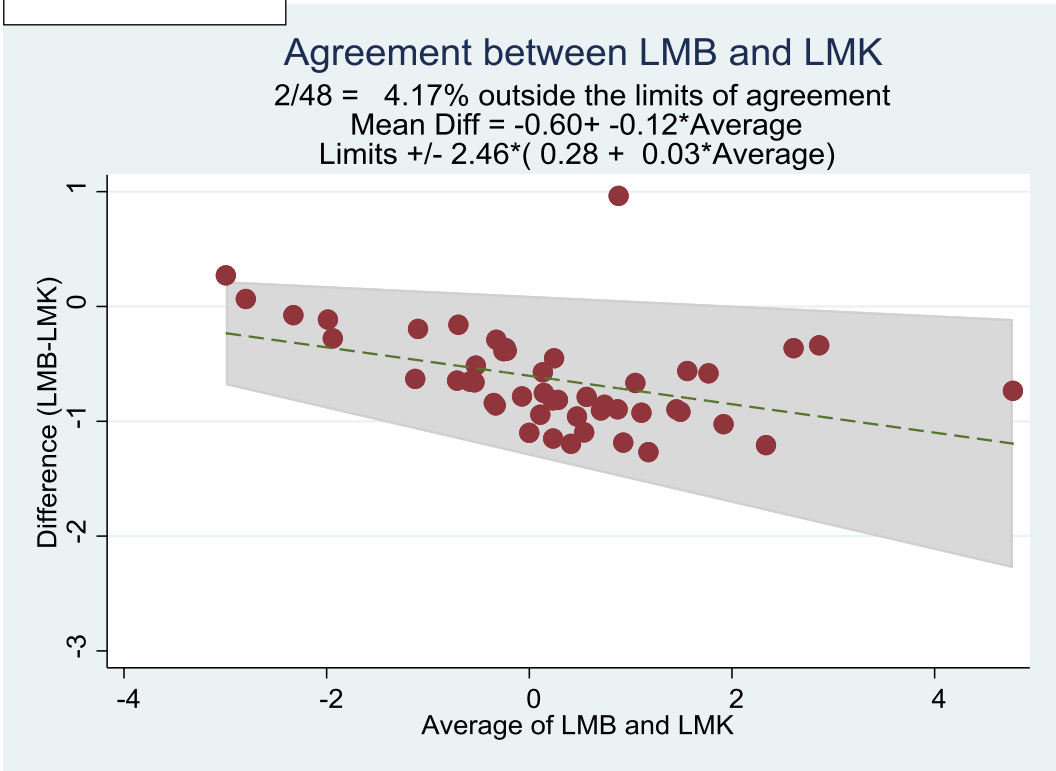


In Misc children, agreement of LMB with LMK

Table 10. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.957[b]	.924	.975	45.135	47	47	.000
Average Measures	.978[d]	.960	.988	45.135	47	47	.000

Fig 9

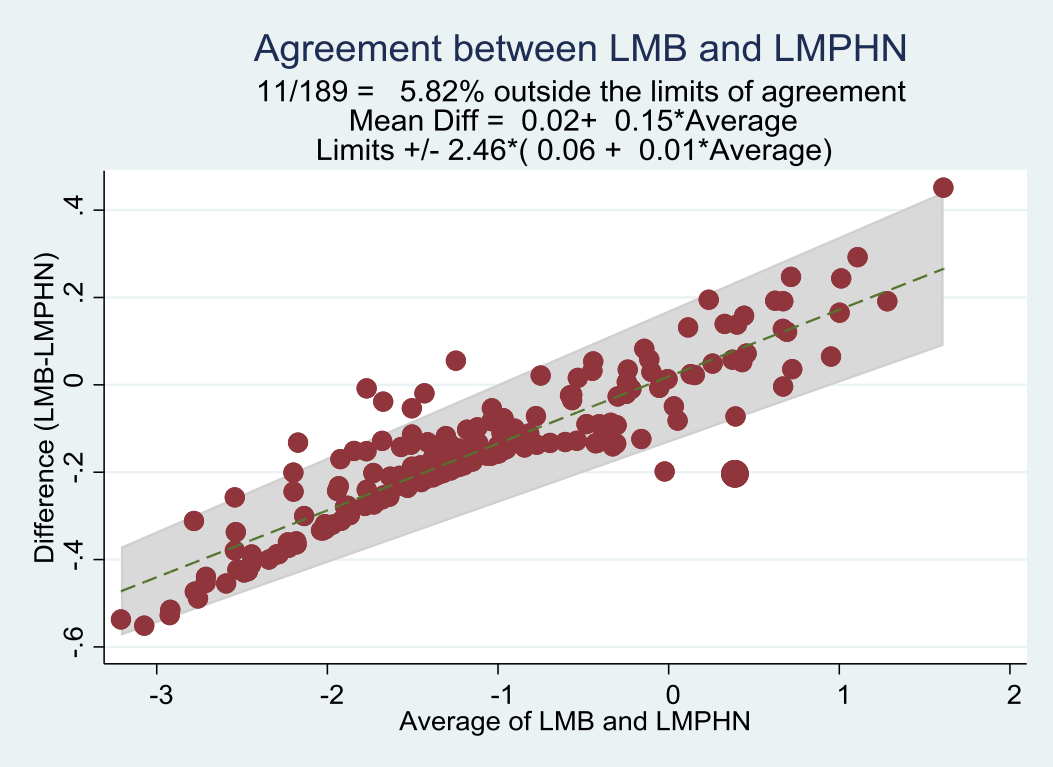


In normal children, agreement of LMB with LMPHN

Table 11. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.986[b]	.981	.989	137.200	188	188	.000
Average Measures	.993[d]	.990	.995	137.200	188	188	.000

Fig 10

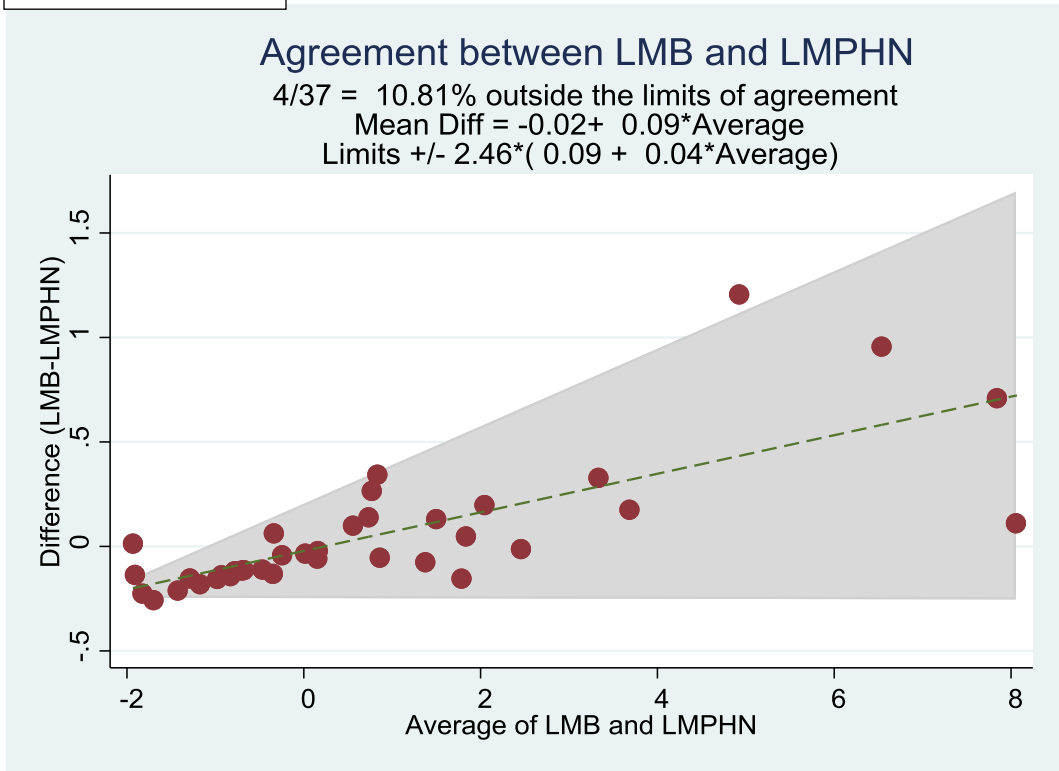


In KD children, agreement of LMB with LMPHN

Table 12. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.992[b]	.985	.996	264.935	36	36	.000
Average Measures	.996[d]	.993	.998	264.935	36	36	.000

Fig 11

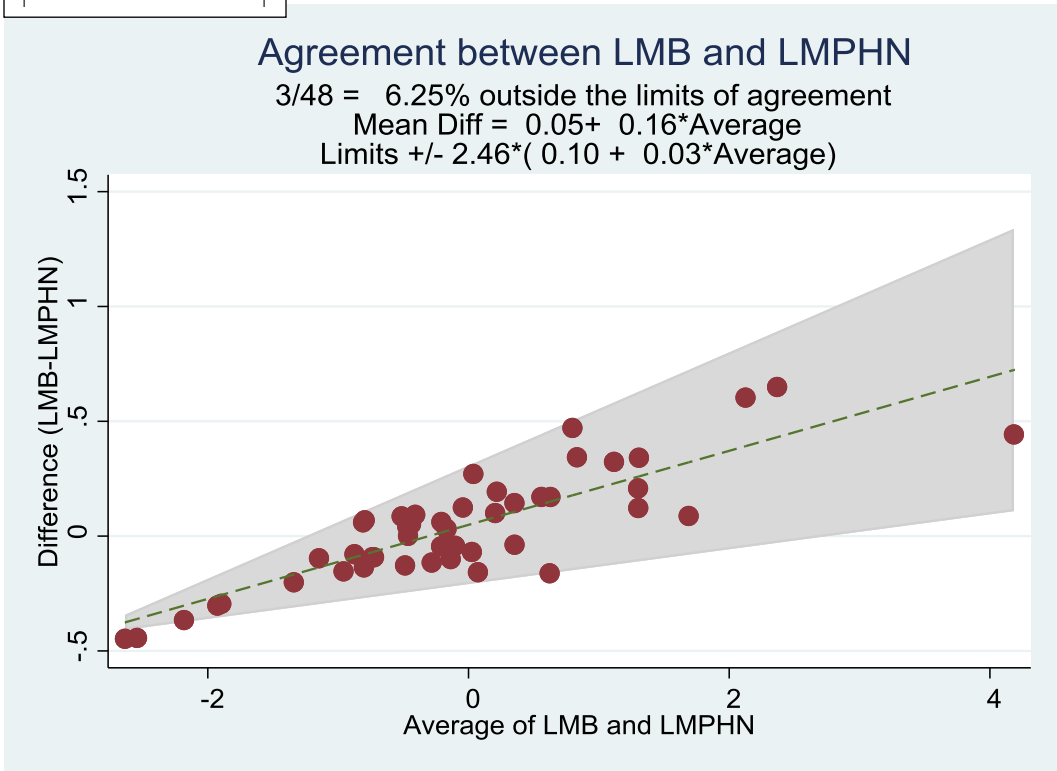


In Misc children, agreement of LMB with LMPHN

Table 12. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.982[b]	.968	.990	109.307	47	47	.000
Average Measures	.991[d]	.984	.995	109.307	47	47	.000

Fig 12

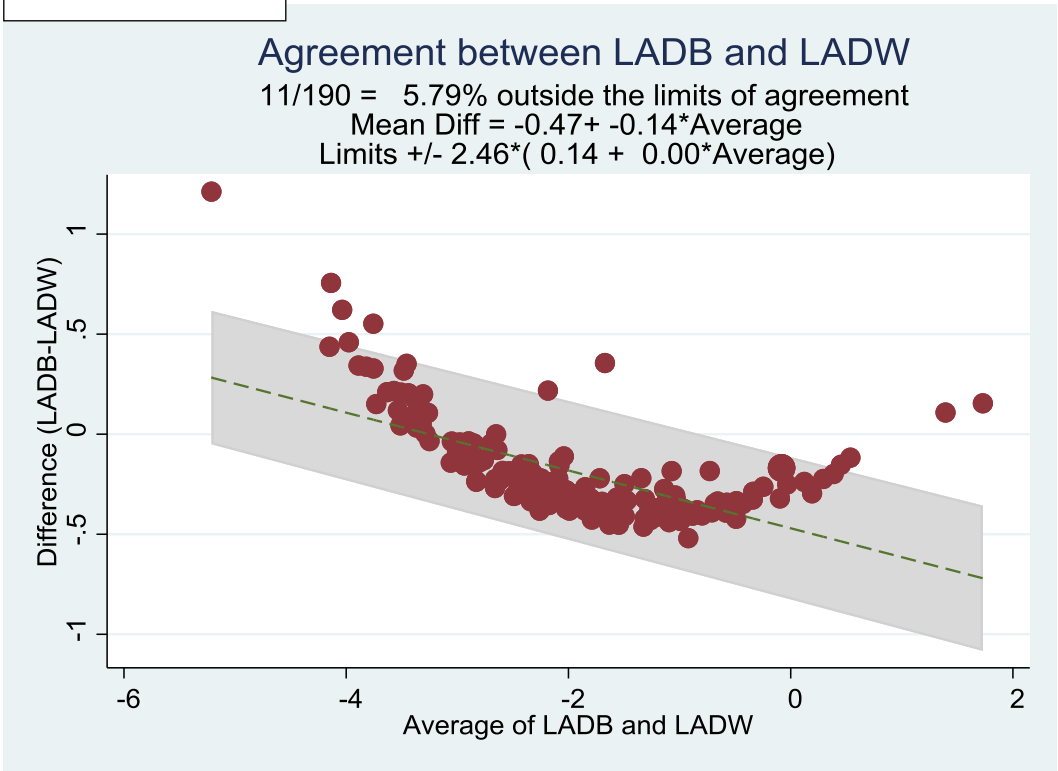


In normal children, agreement of LADB with LADW

Table 13. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.975[b]	.967	.981	78.238	189	189	.000
Average Measures	.987[d]	.983	.990	78.238	189	189	.000

Fig 13.

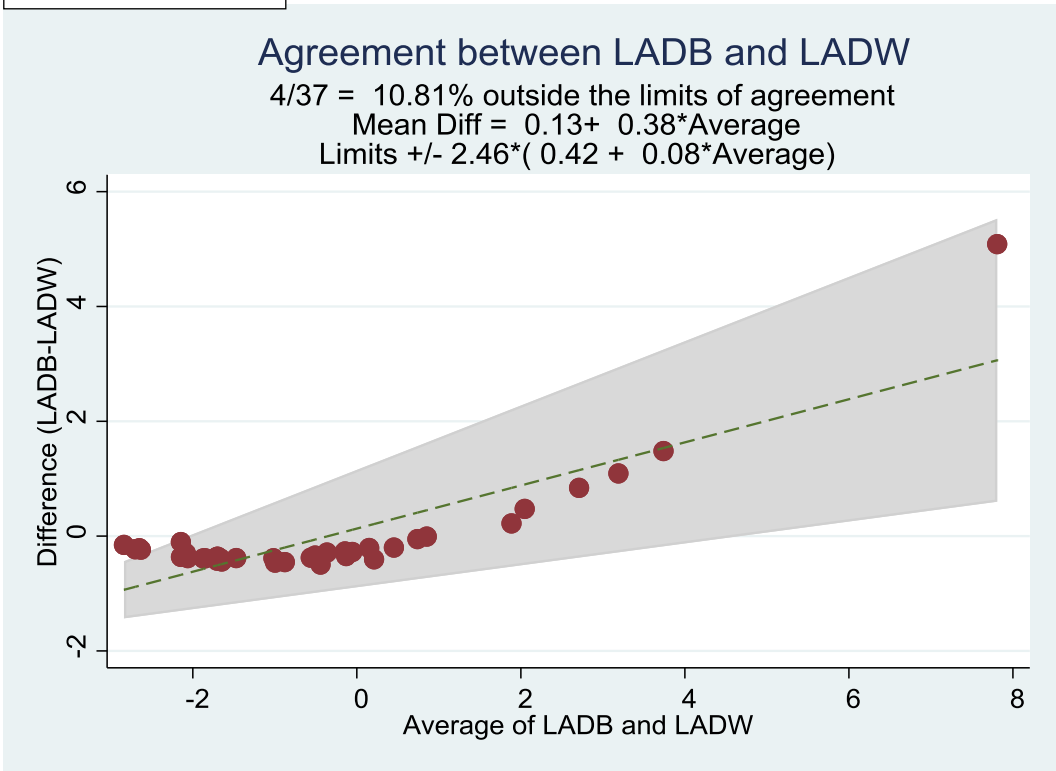


In KD children, agreement of LADB with LADW

Table 14. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.906[b]	.825	.950	20.235	36	36	.000
Average Measures	.951[d]	.904	.975	20.235	36	36	.000

Fig 14

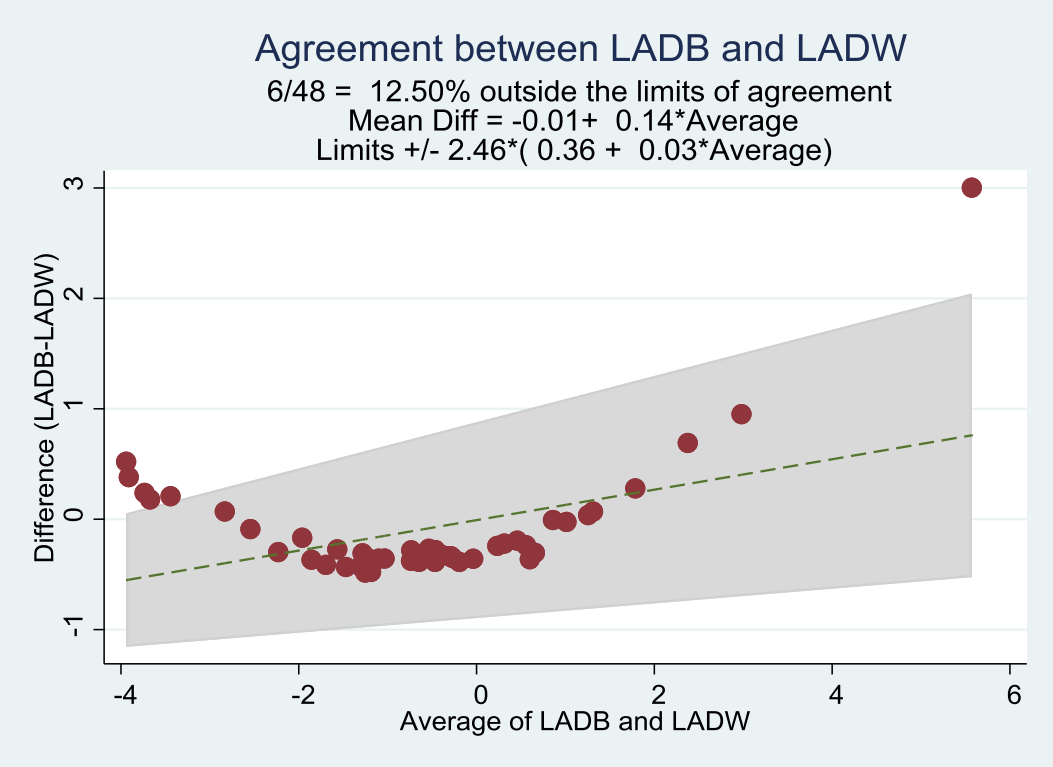


In Misc children, agreement of LADB with LADW

Table 15. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.954[b]	.920	.974	42.852	47	47	.000
Average Measures	.977[d]	.958	.987	42.852	47	47	.000

Fig 15

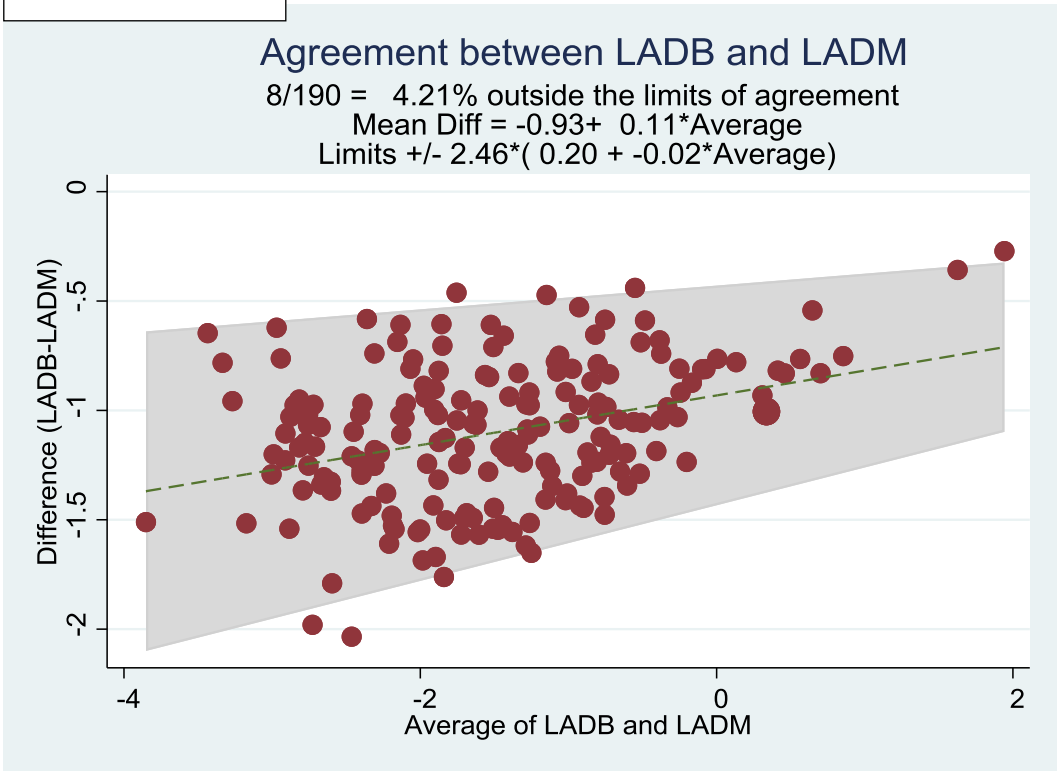


In normal children, agreement of LADB with LADM

Table 16. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.949[b]	.933	.961	38.257	189	189	.000
Average Measures	.974[d]	.965	.980	38.257	189	189	.000

Fig 16

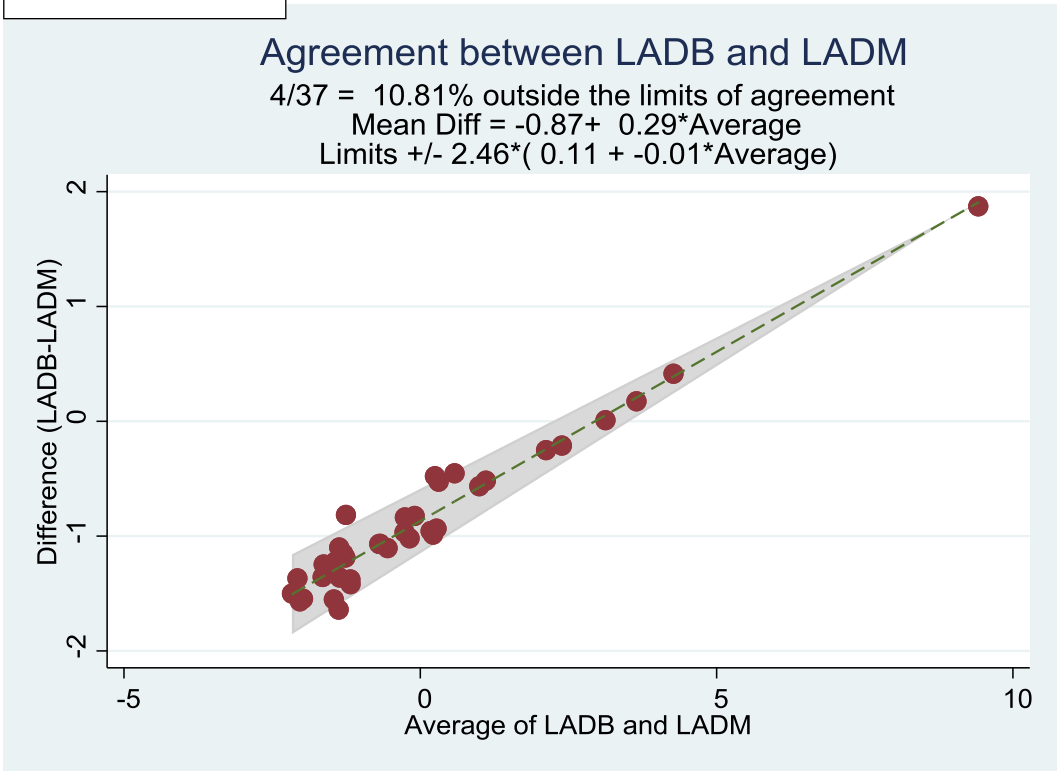


In KD children, agreement of LADB with LADM

Table 17. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.955[b]	.915	.977	43.655	36	36	.000
Average Measures	.977[d]	.956	.988	43.655	36	36	.000

Fig 17

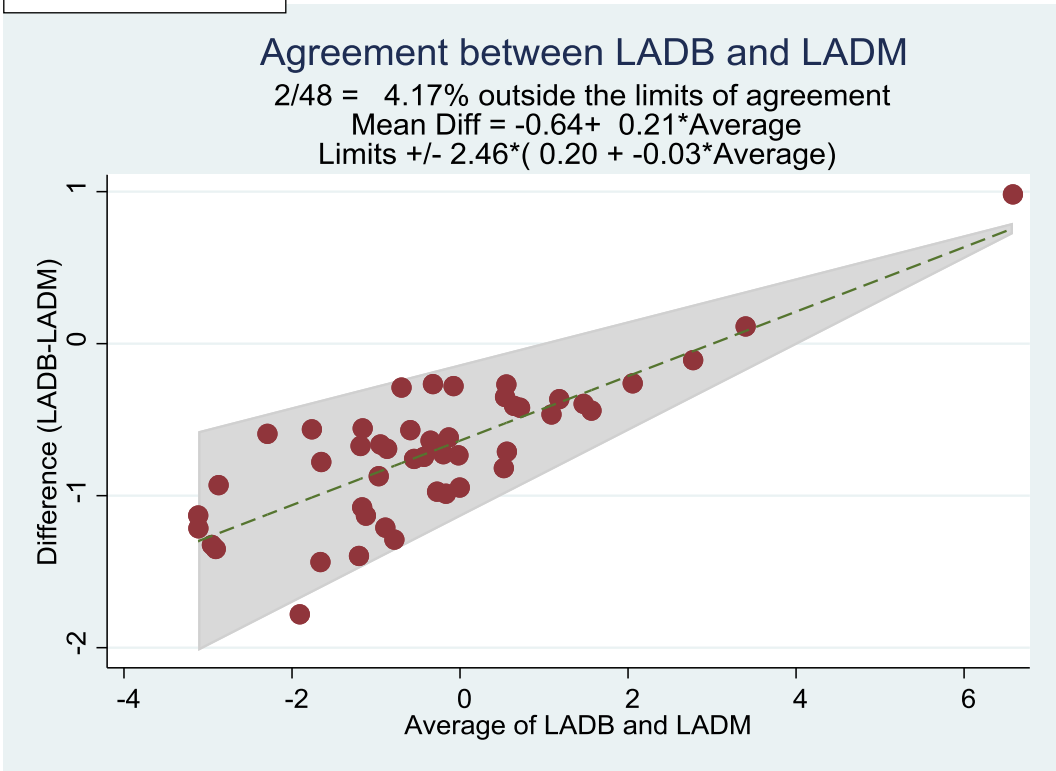


In Misc children, agreement of LADB with LADM

Table 18. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.966[b]	.940	.981	57.567	47	47	.000
Average Measures	.983[d]	.969	.990	57.567	47	47	.000

Fig 18

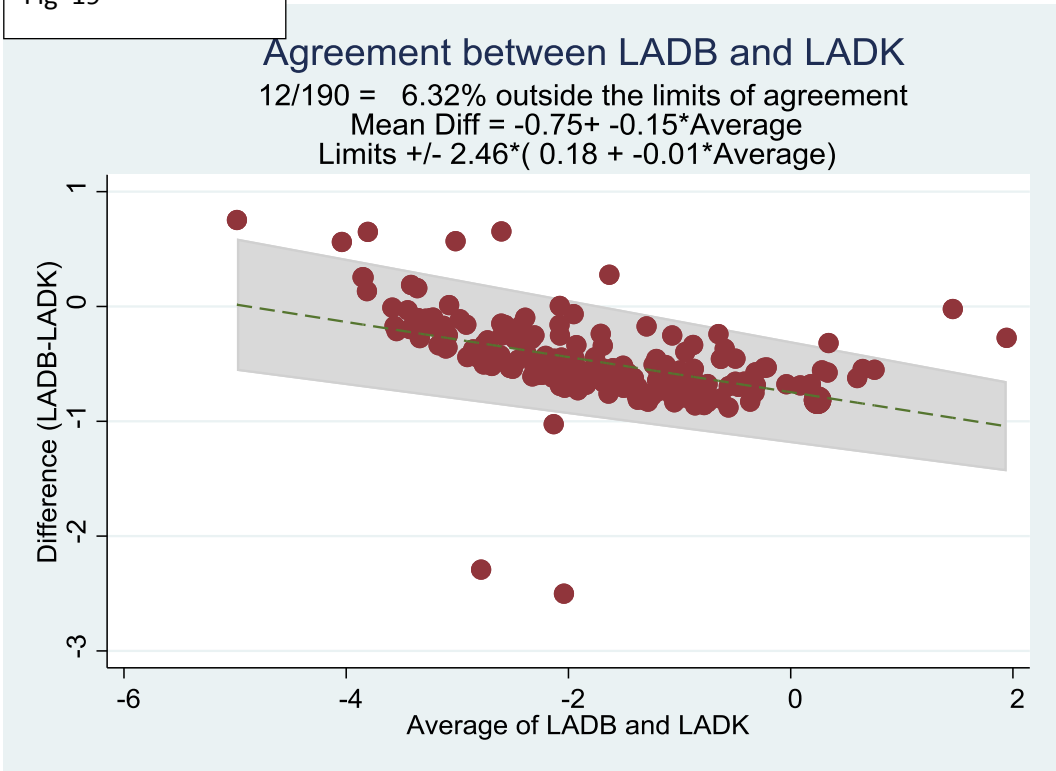


In normal children, agreement of LADB with LADK

Table 19. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.949[b]	.932	.961	37.981	189	189	.000
Average Measures	.974[d]	.965	.980	37.981	189	189	.000

Fig 19

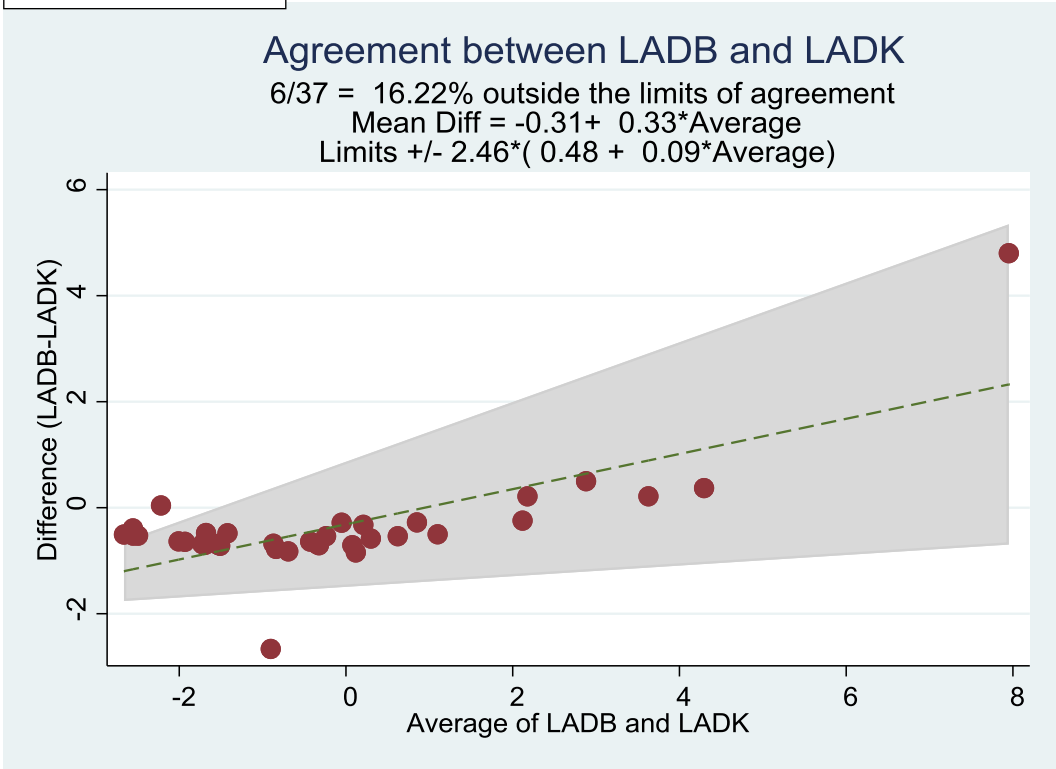


In KD children, agreement of LADB with LADK

Table 20. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.902[b]	.818	.948	19.347	36	36	.000
Average Measures	.948[d]	.900	.973	19.347	36	36	.000

Fig 20

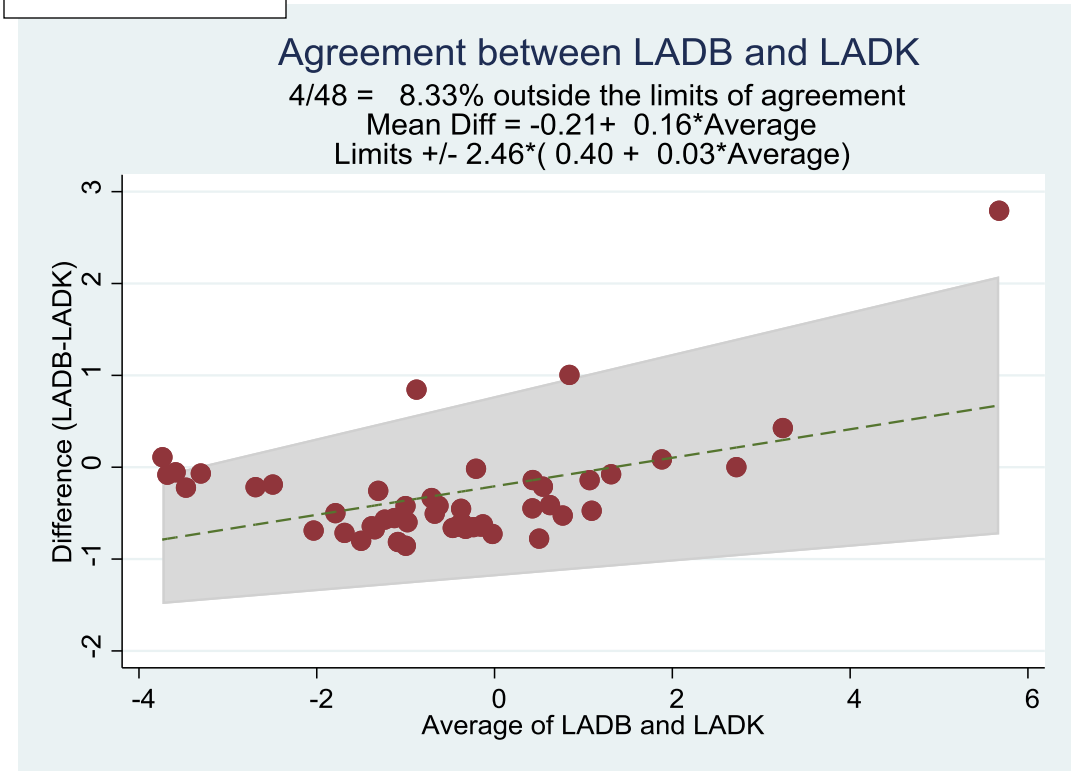


In Misc children, agreement of LADB with LADK

Table 21. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.946[b]	.905	.969	35.771	47	47	.000
Average Measures	.972[d]	.950	.984	35.771	47	47	.000

Fig 21

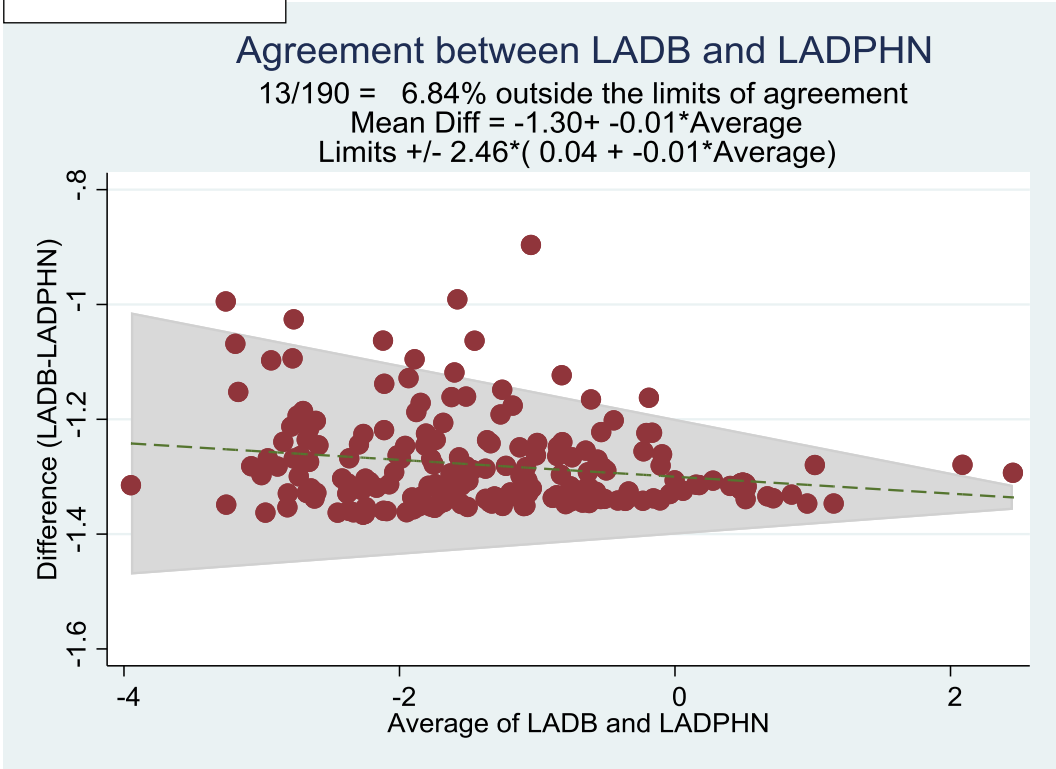


In normal children, agreement of LADB with LADPHN

Table 22. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.997[b]	.996	.998	680.497	189	189	.000
Average Measures	.999[d]	.998	.999	680.497	189	189	.000

Fig 22

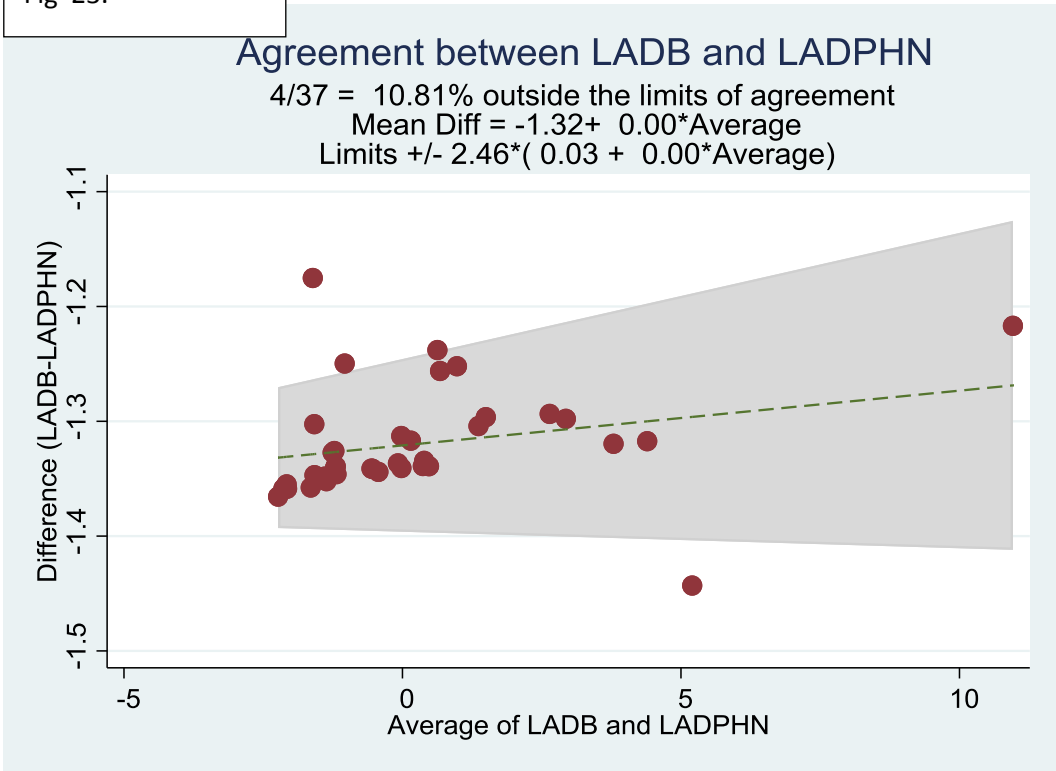


In KD children, agreement of LADB with LADPHN

Table 23. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	1.000[b]	1.000	1.000	11452.687	36	36	.000
Average Measures	1.000[d]	1.000	1.000	11452.687	36	36	.000

Fig 23.

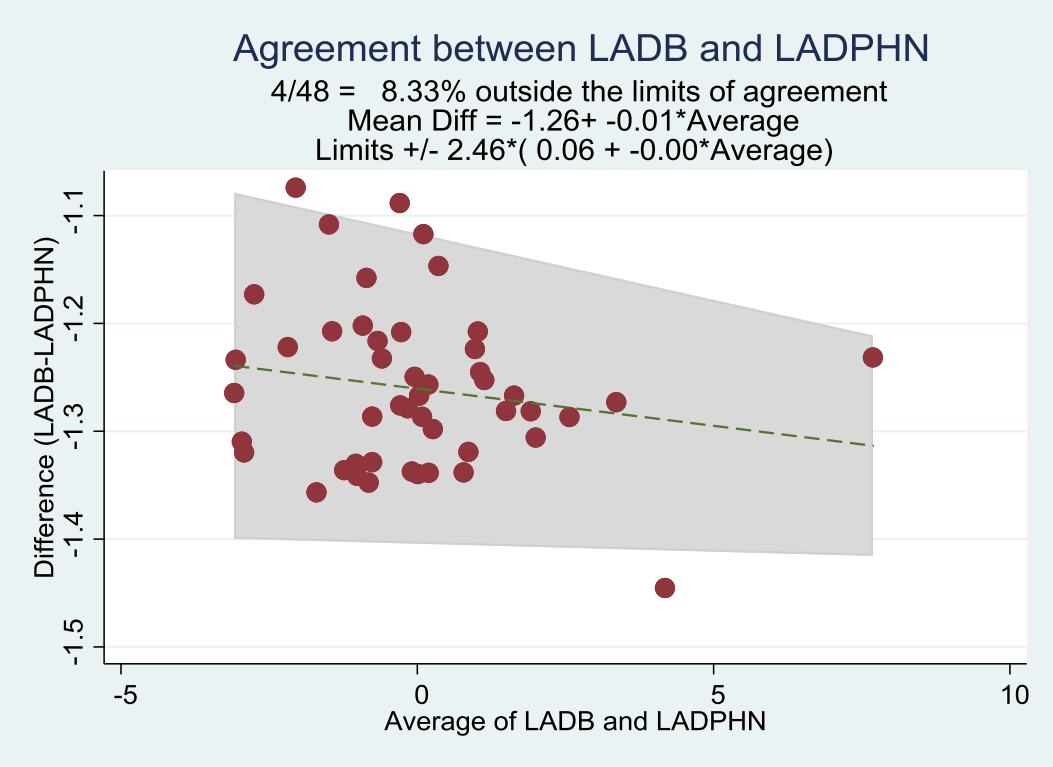


In Misc children, agreement of LADB with LADPHN

Table 24. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.999[b]	.999	1.000	2660.769	47	47	.000
Average Measures	1.000[d]	.999	1.000	2660.769	47	47	.000

Fig 24

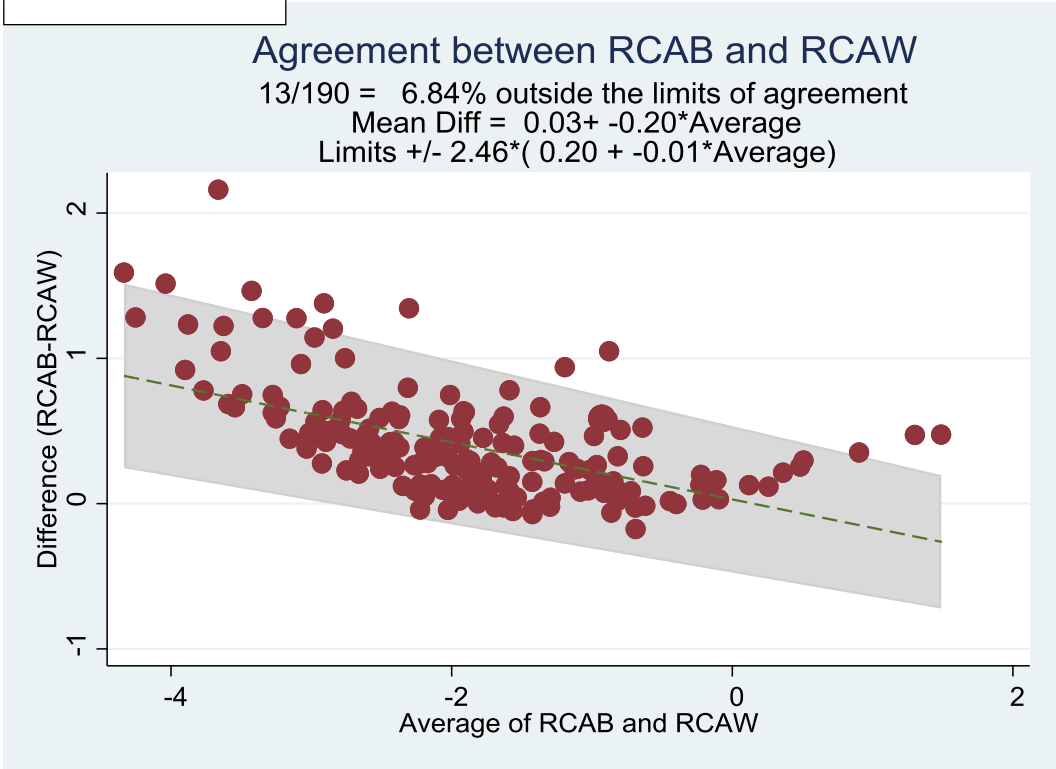


In normal children, agreement of RCAB with RCAW

Table 25. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.941[b]	.922	.955	32.633	189	189	.000
Average Measures	.969[d]	.959	.977	32.633	189	189	.000

Fig 25

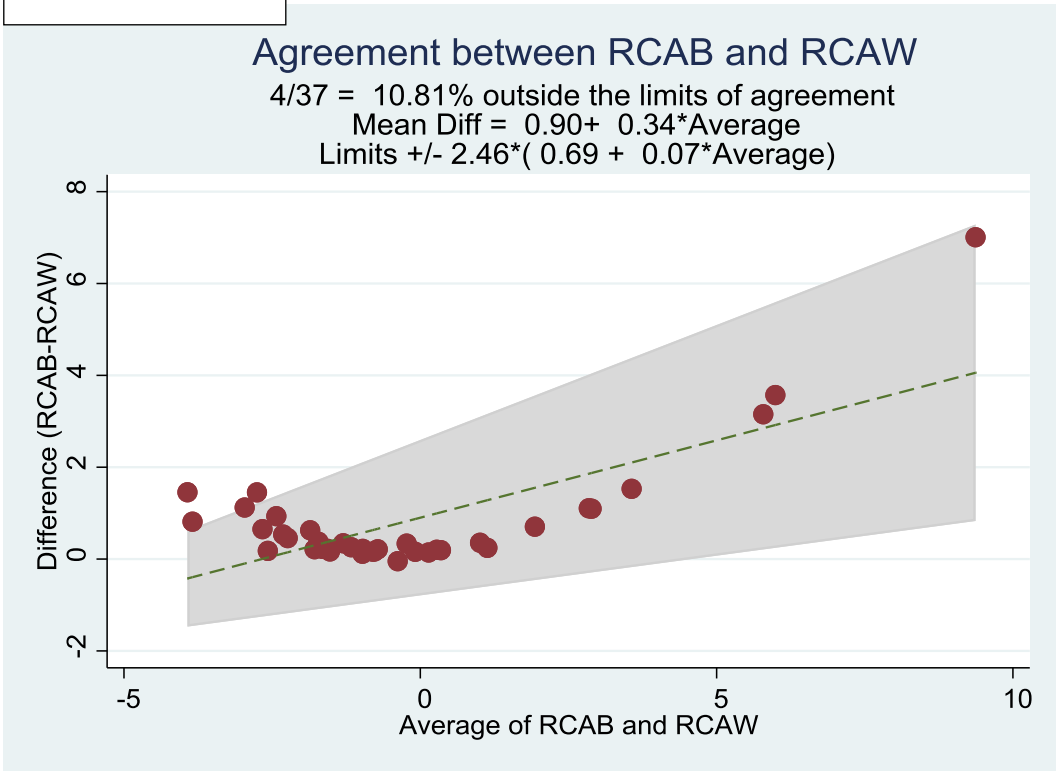


In KD children, agreement of RCAB with RCAW

Table 26. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.900[b]	.815	.947	19.082	36	36	.000
Average Measures	.948[d]	.898	.973	19.082	36	36	.000

Fig 26

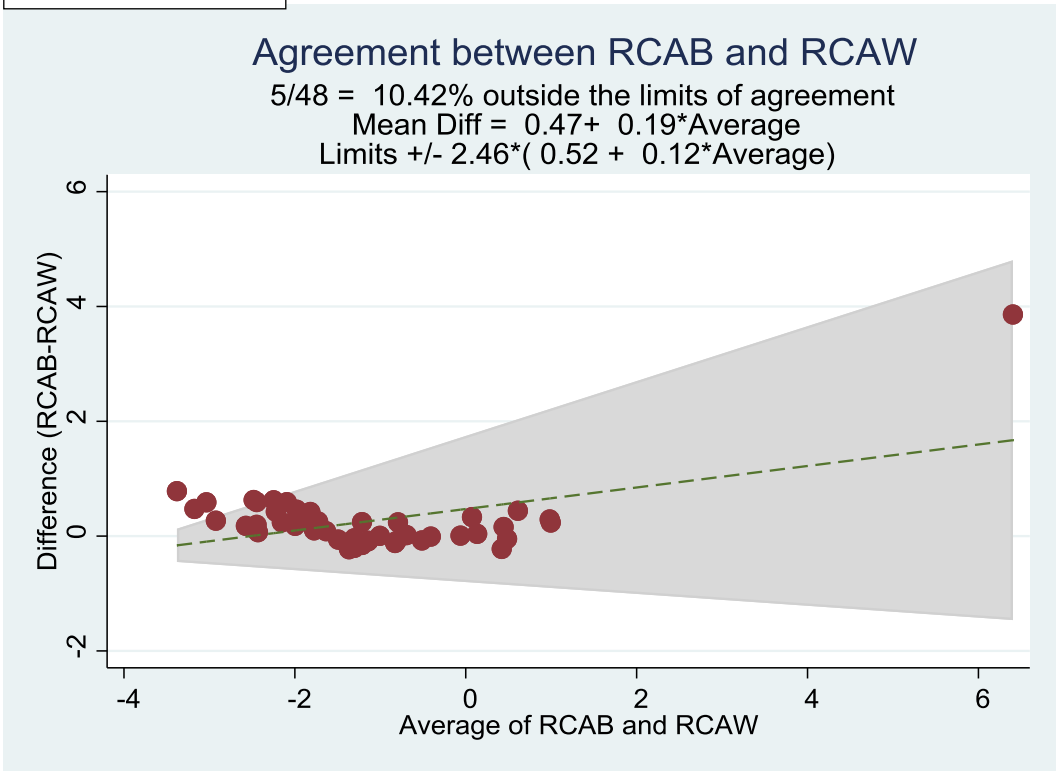


In Misc children, agreement of RCAB with RCAW

Table 27. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.932[b]	.882	.961	28.404	47	47	.000
Average Measures	.965[d]	.937	.980	28.404	47	47	.000

Fig 27

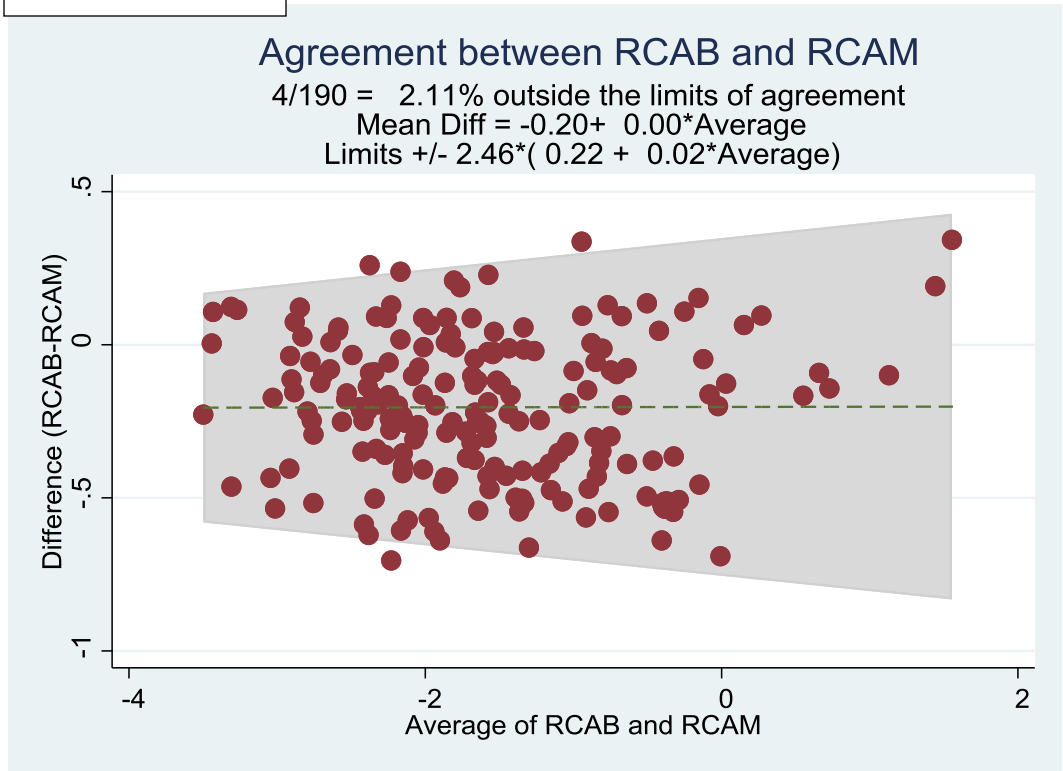


In normal children, agreement of RCAB with RCAM

Table 28. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.971[b]	.962	.978	67.922	189	189	.000
Average Measures	.985[d]	.980	.989	67.922	189	189	.000

Fig 28

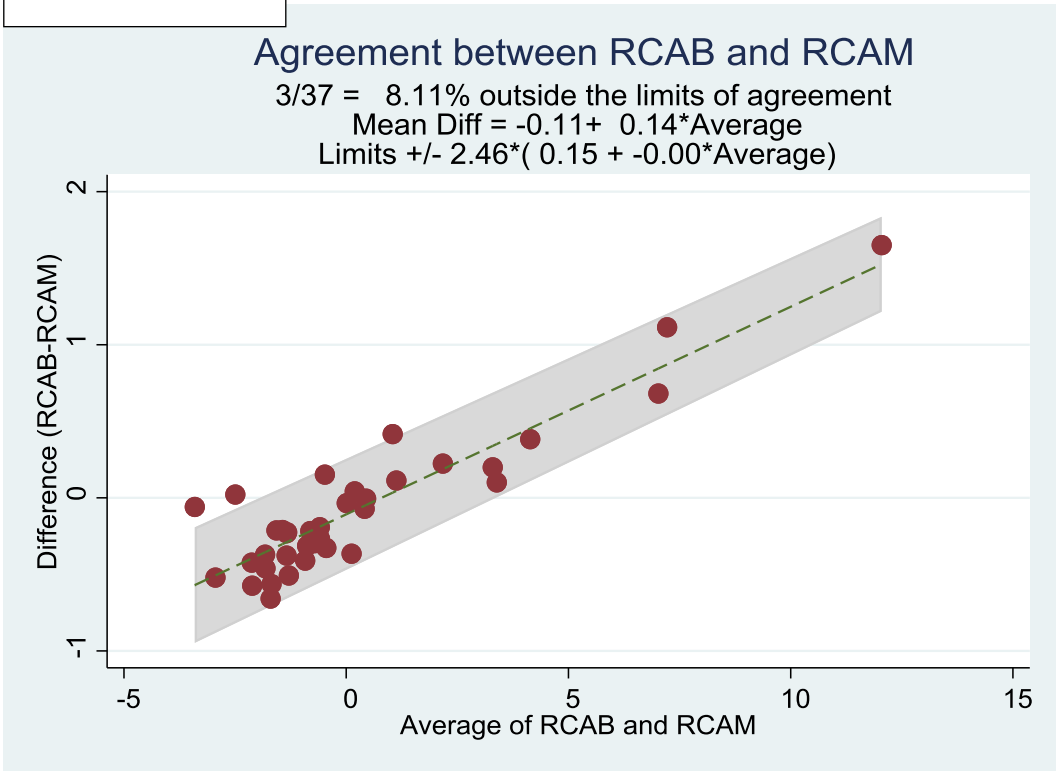


In KD children, agreement of RCAB with RCAM

Table 29. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.989[b]	.979	.994	179.391	36	36	.000
Average Measures	.994[d]	.989	.997	179.391	36	36	.000

Fig 29

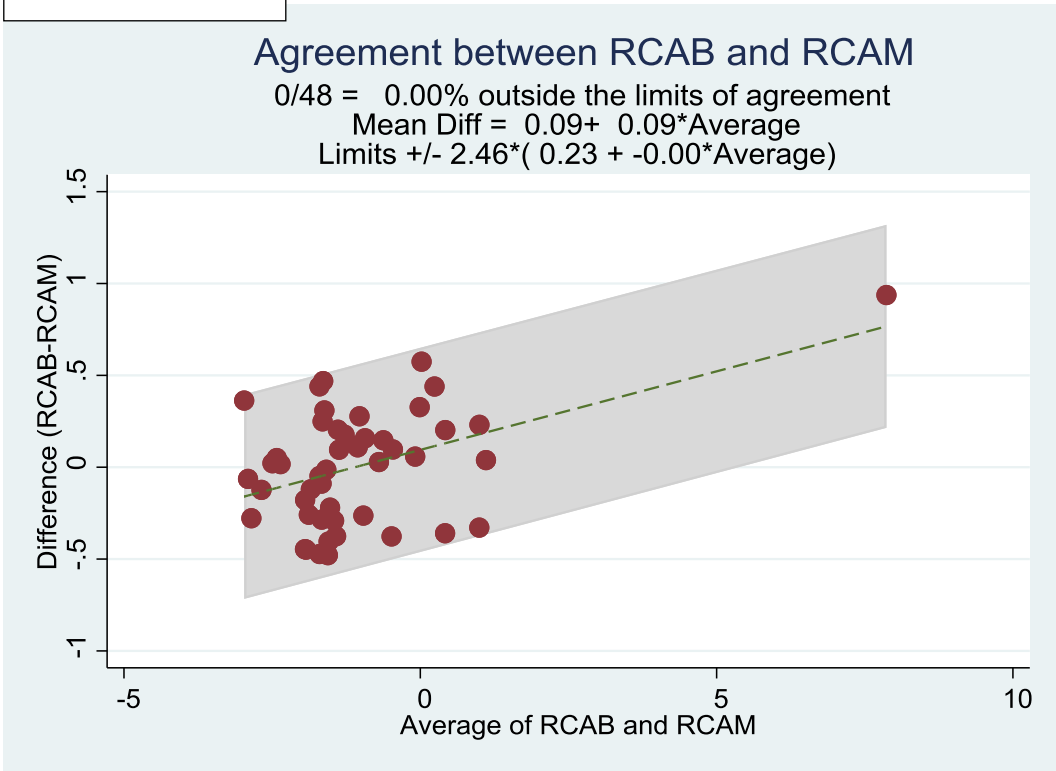


In Misc children, agreement of RCAB with RCAM

Table 30. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.982[b]	.969	.990	112.820	47	47	.000
Average Measures	.991[d]	.984	.995	112.820	47	47	.000

Fig 30

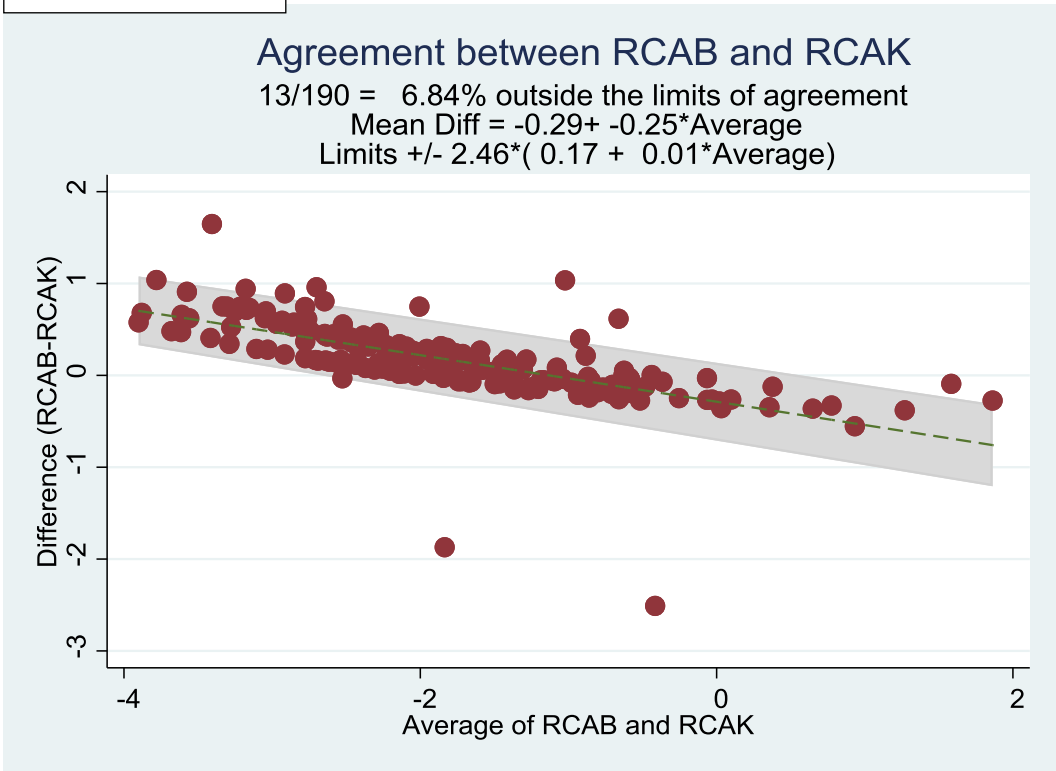


In normal children, agreement of RCAB with RCAK

Table 31. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.930[b]	.908	.947	27.633	189	189	.000
Average Measures	.964[d]	.952	.973	27.633	189	189	.000

Fig 31

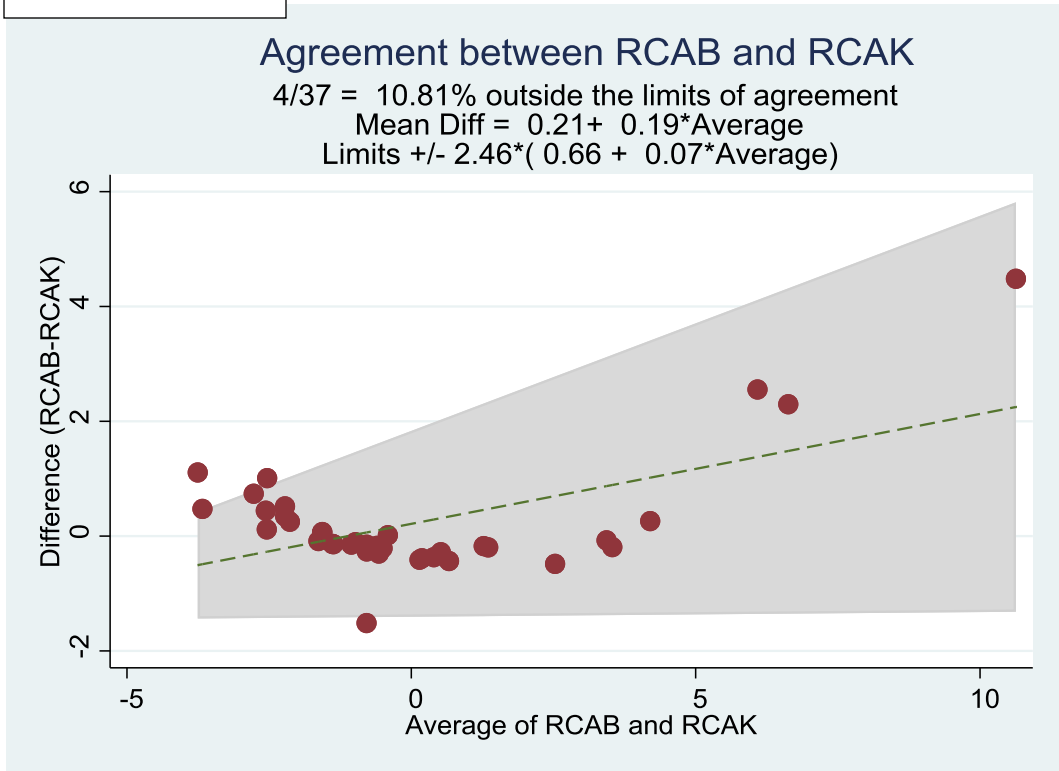


In KD children, agreement of RCAB with RCAK

Table 32. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.945[b]	.896	.971	35.516	36	36	.000
Average Measures	.972[d]	.945	.986	35.516	36	36	.000

Fig 32

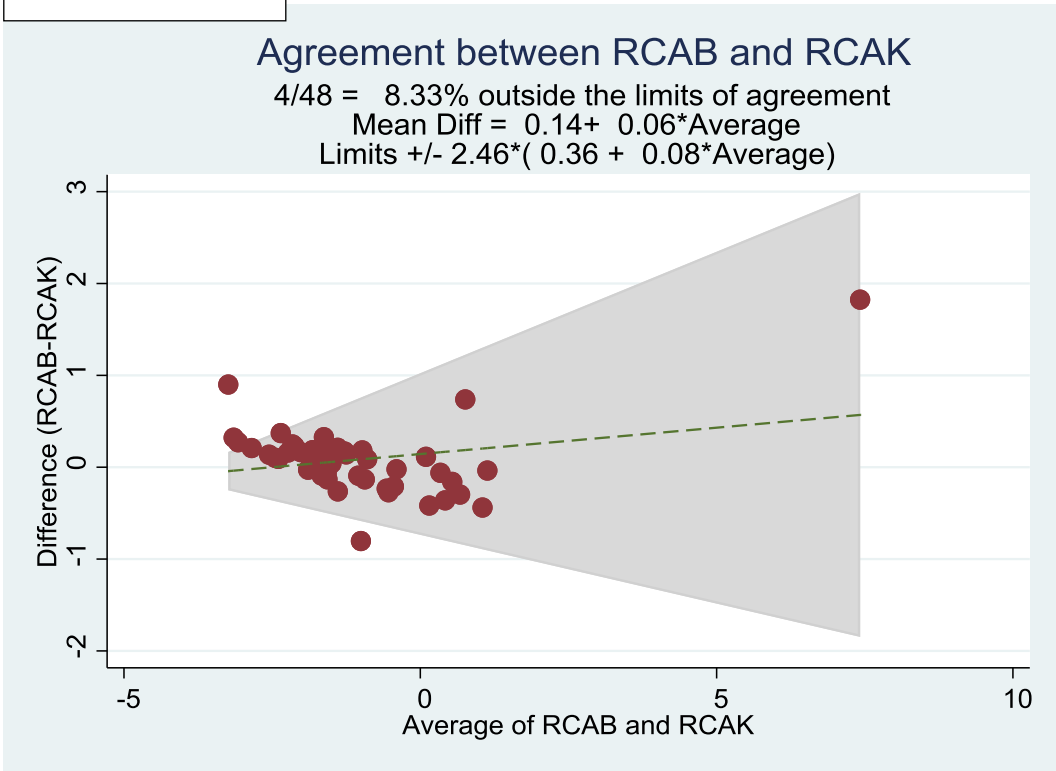


In Misc children, agreement of RCAB with RCAK

Table 33. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.974[b]	.955	.986	77.022	47	47	.000
Average Measures	.987[d]	.977	.993	77.022	47	47	.000

Fig 33

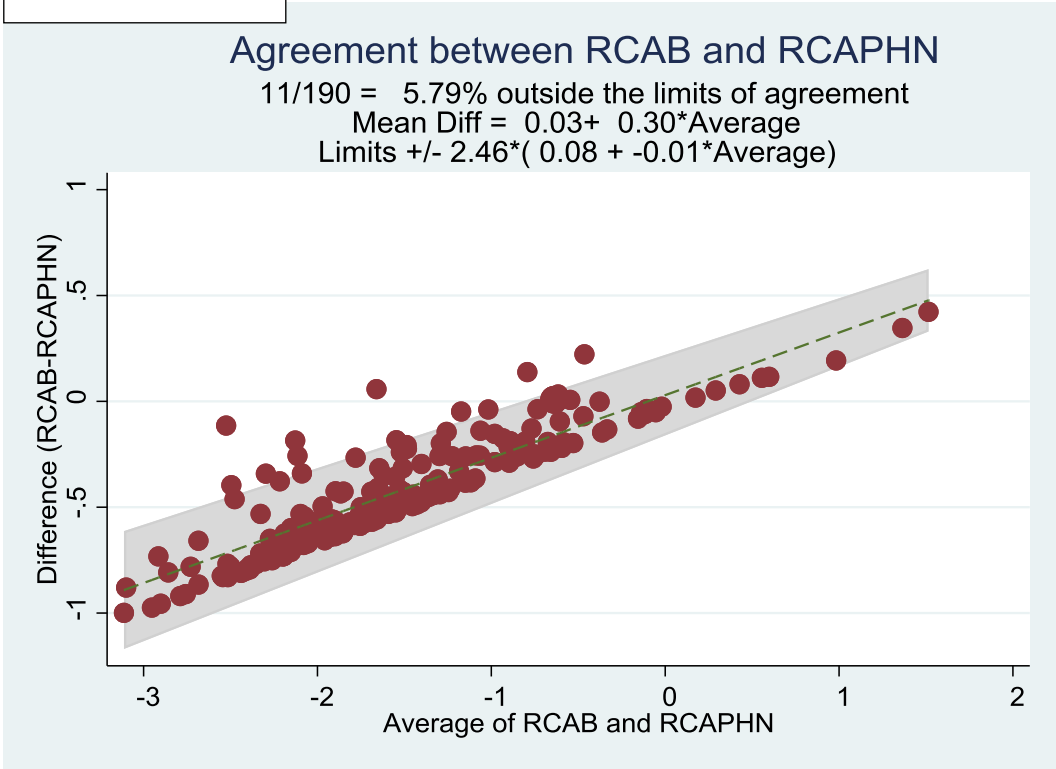


In normal children, agreement of RCAB with RCAPHN

Table 34. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.946[b]	.929	.959	36.195	189	189	.000
Average Measures	.972[d]	.963	.979	36.195	189	189	.000

Fig 34

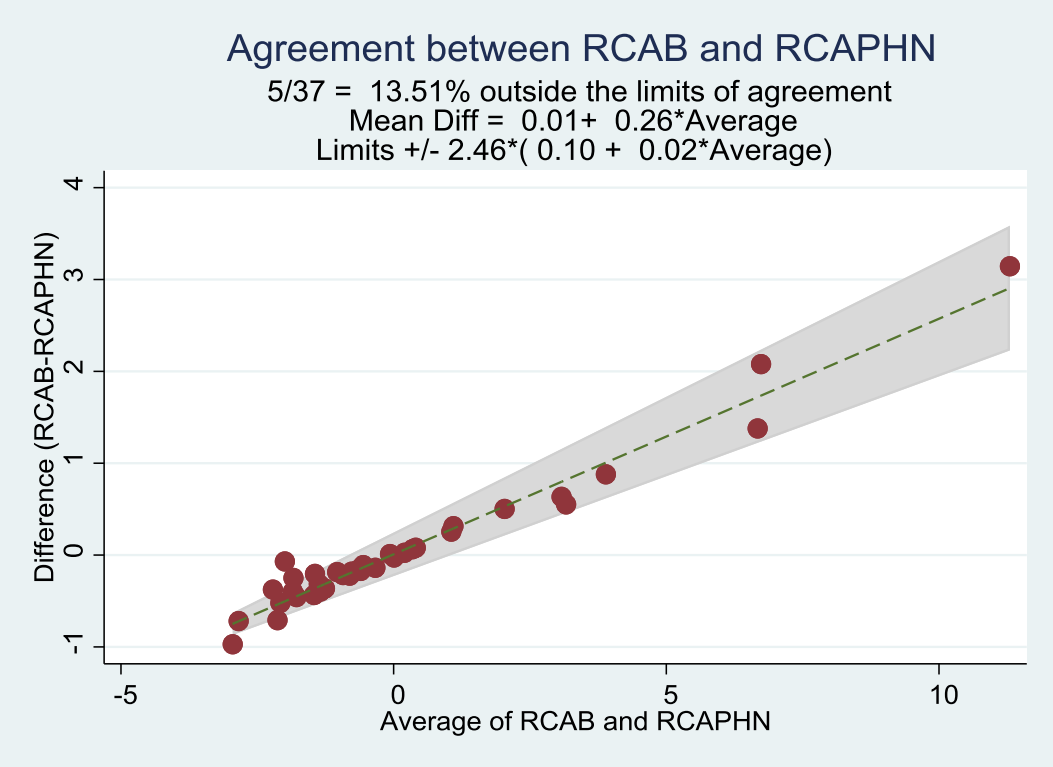


In KD children, agreement of RCAB with RCAPHN

Table 35. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.966[b]	.936	.983	58.528	36	36	.000
Average Measures	.983[d]	.967	.991	58.528	36	36	.000

Fig 35

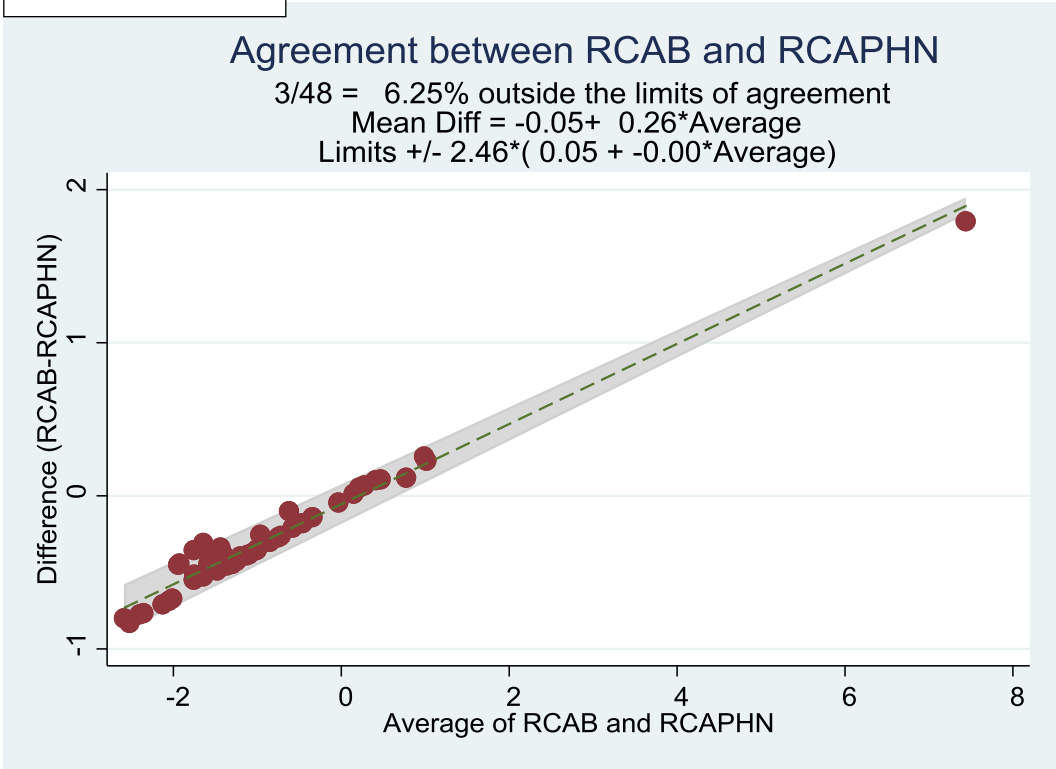


In Misc children, agreement of RCAB with RCAPHN

Table 36. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.965[b]	.939	.980	56.628	47	47	.000
Average Measures	.982[d]	.968	.990	56.628	47	47	.000

Fig 36

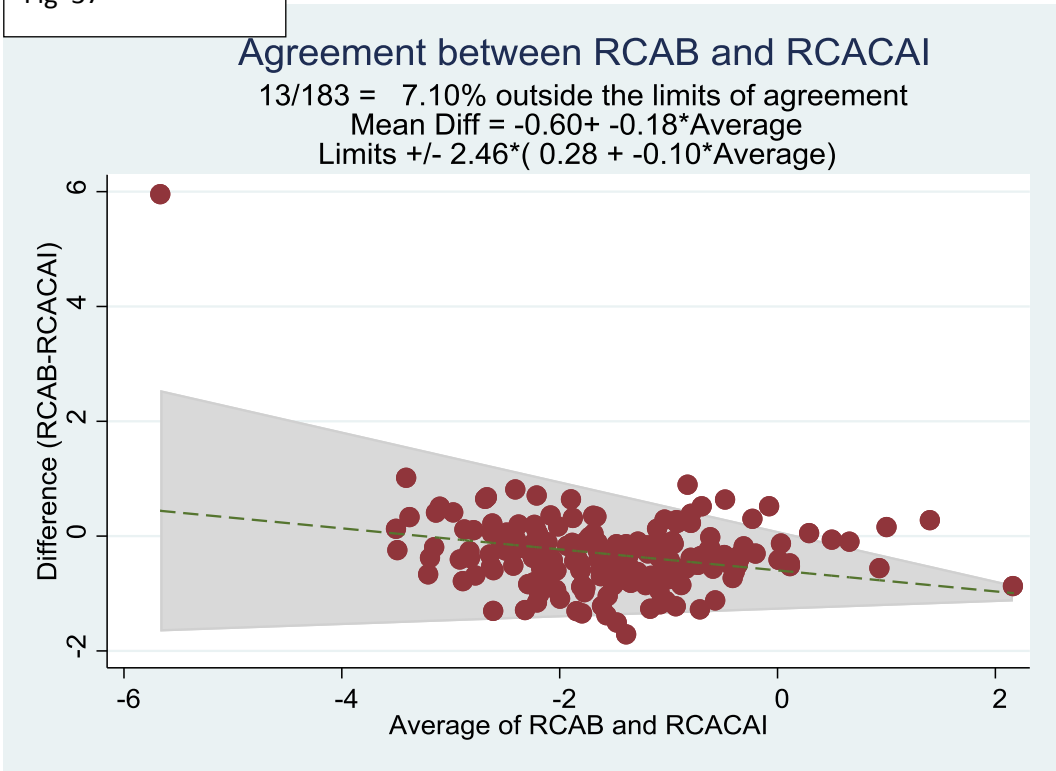


In normal children, agreement of RCAB with RCACAI

Table 37. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.791[b]	.730	.840	8.566	182	182	.000
Average Measures	.883[d]	.844	.913	8.566	182	182	.000

Fig 37

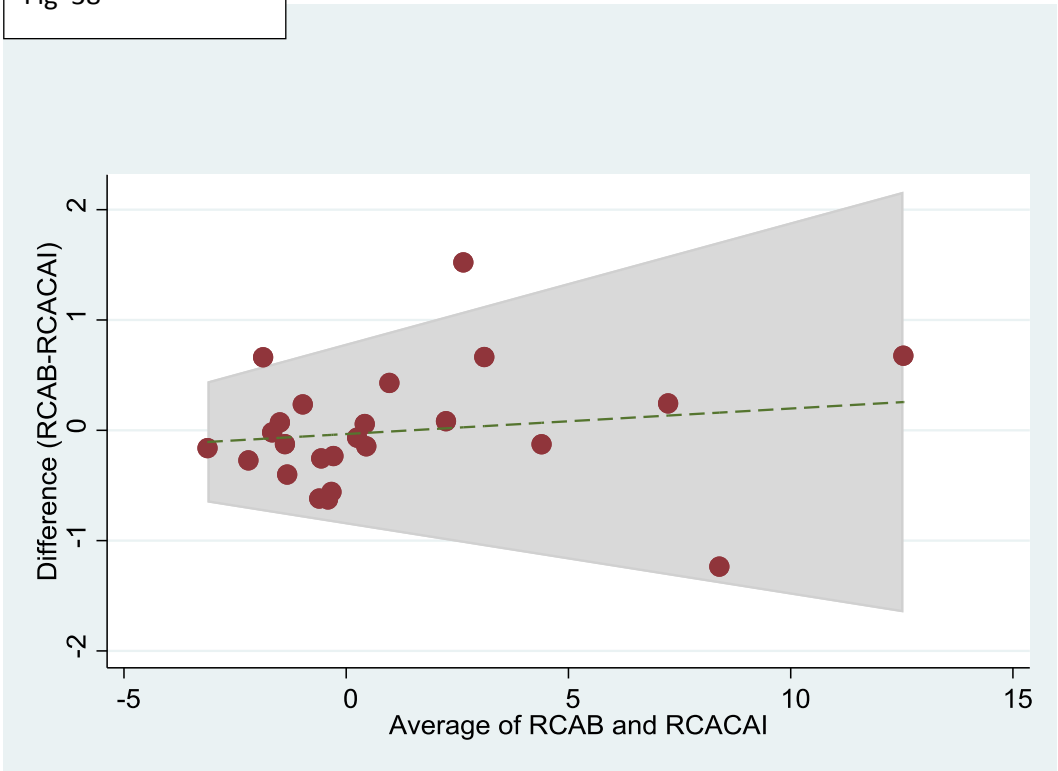


In KD children, agreement of RCAB with RCACAI

Table 38. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.989[b]	.975	.995	183.534	23	23	.000
Average Measures	.995[d]	.987	.998	183.534	23	23	.000

Fig 38

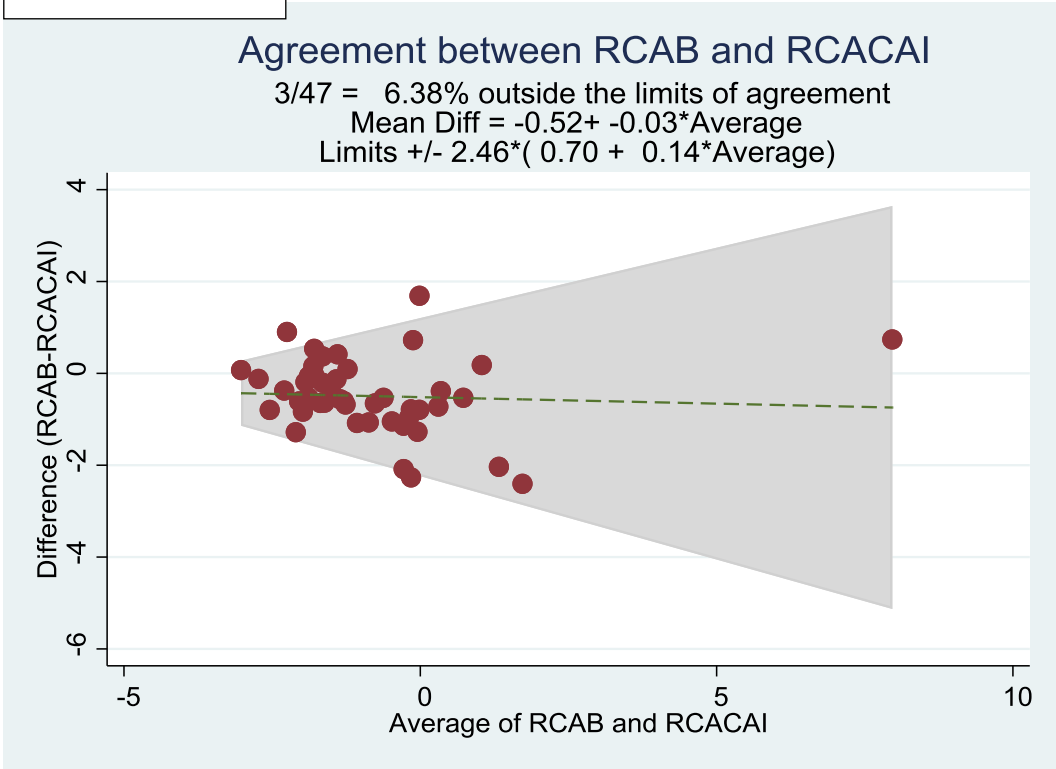


In Misc children, agreement of RCAB with RCACAI

Table 39. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.894[b]	.817	.940	17.871	46	46	.000
Average Measures	.944[d]	.900	.969	17.871	46	46	.000

Fig 39

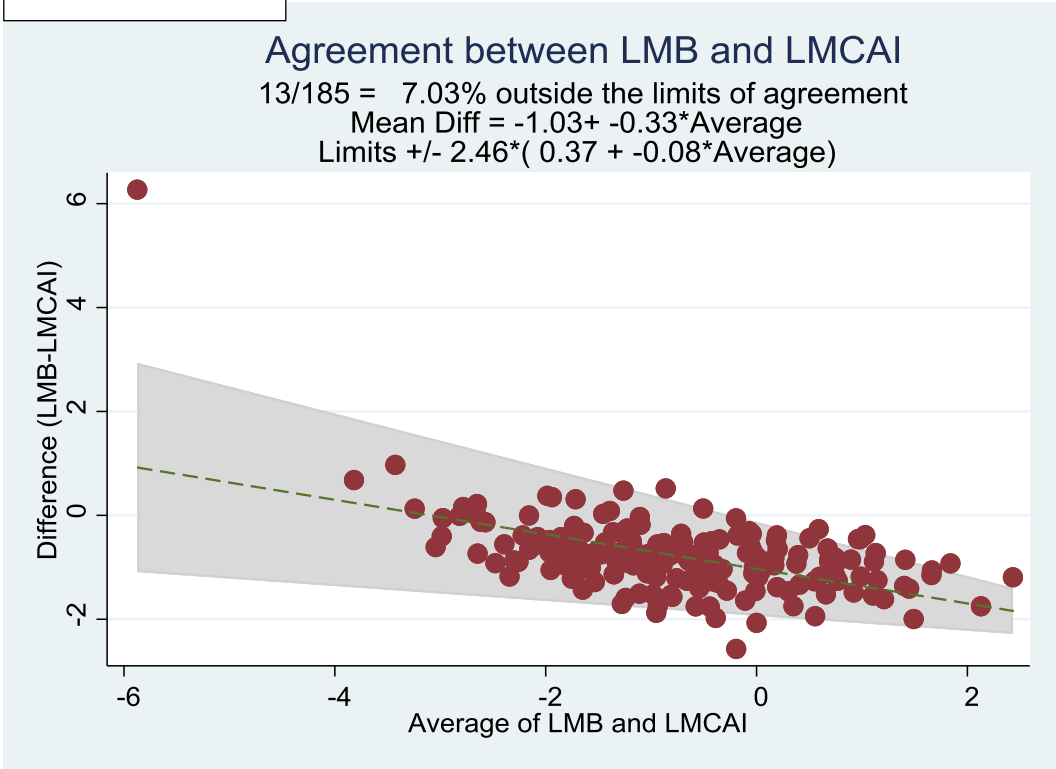


In normal children, agreement of LMB with LMCAI

Table 40. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.826[b]	.774	.867	10.513	184	184	.000
Average Measures	.905[d]	.873	.929	10.513	184	184	.000

Fig 40

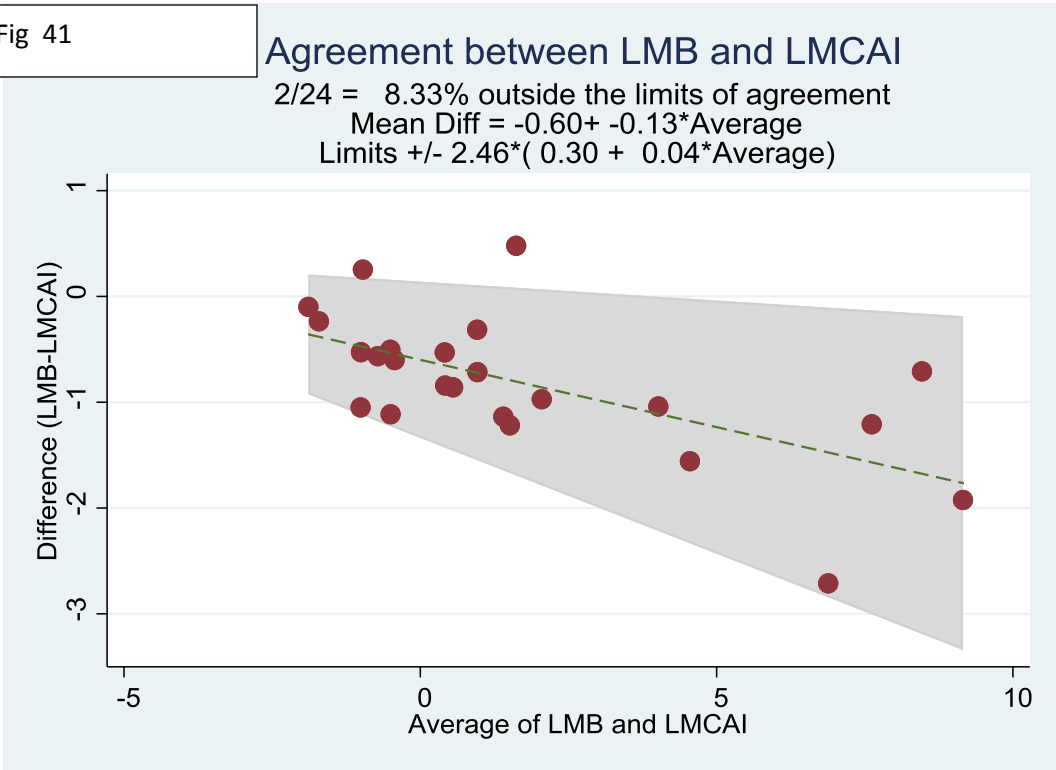


In KD children, agreement of LMB with LMCAI

Table 41. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.979[b]	.953	.991	96.423	23	23	.000
Average Measures	.990[d]	.976	.996	96.423	23	23	.000

Fig 41

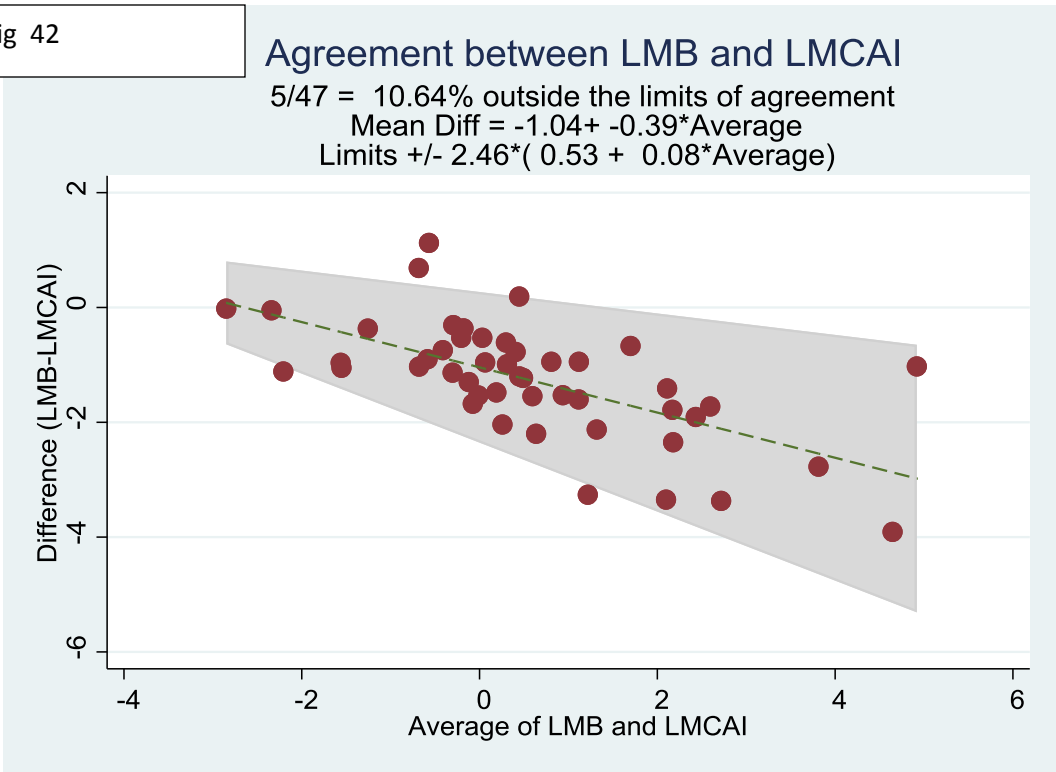


In Misc children, agreement of LMB with LMCAI

Table 42. Intraclass Correlation Coefficient[a]

	Intraclass Correlation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.825[b]	.706	.898	10.414	46	46	.000
Average Measures	.904[d]	.828	.947	10.414	46	46	.000

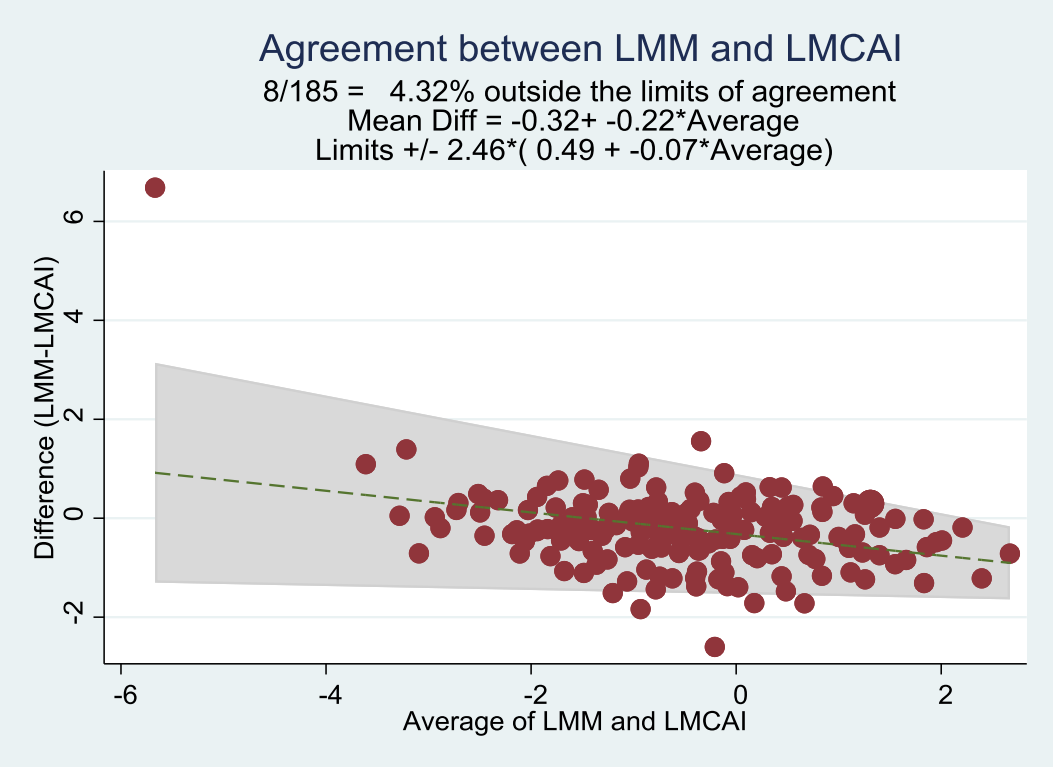
Fig 42



In normal children, agreement between LMM and LMCAI

Intraclass Correlation Coefficient[a]

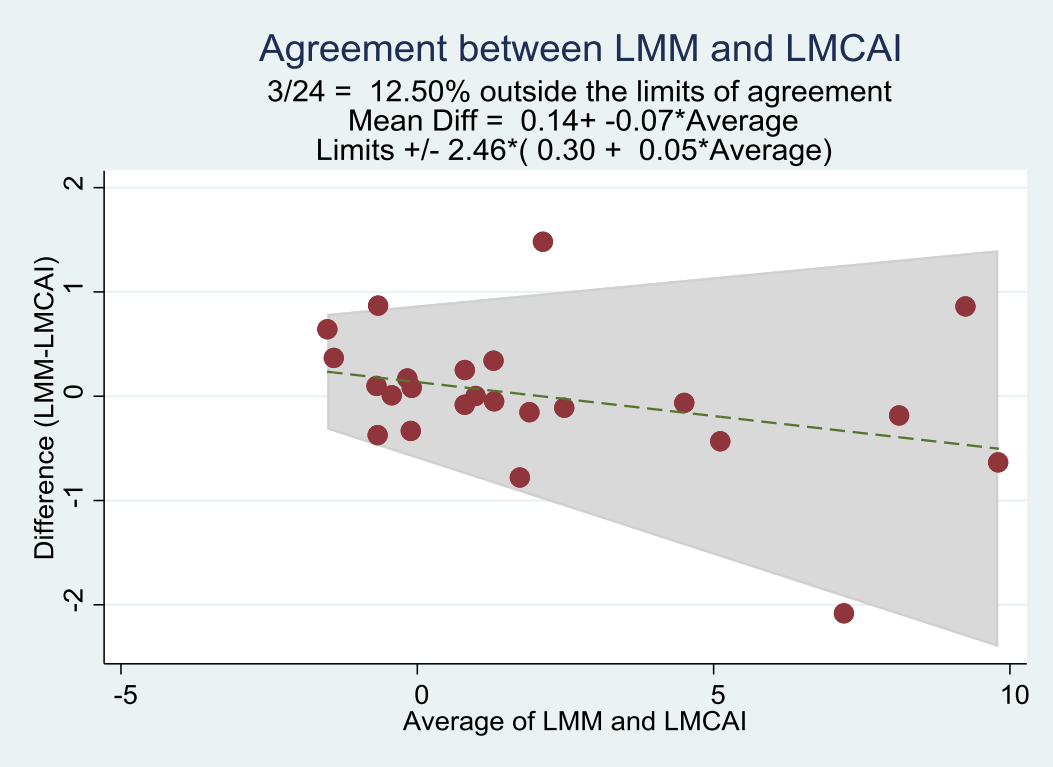
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.818[b]	.764	.861	9.982	184	184	.000
Average Measures	.900[d]	.866	.925	9.982	184	184	.000



In KD children, agreement between LMM and LMCAI

Intraclass Correlation Coefficient[a]

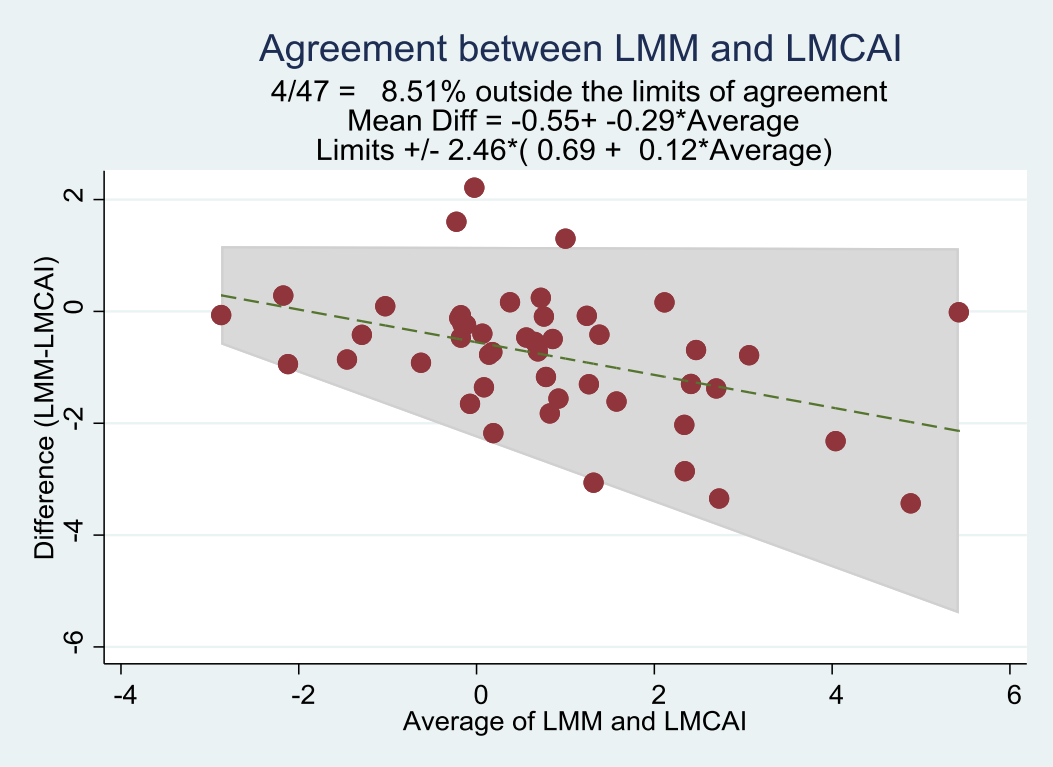
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.981[b]	.956	.992	102.090	23	23	.000
Average Measures	.990[d]	.977	.996	102.090	23	23	.000



In Misc children, agreement between LMM and LMCAI

Intraclass Correlation Coefficient[a]

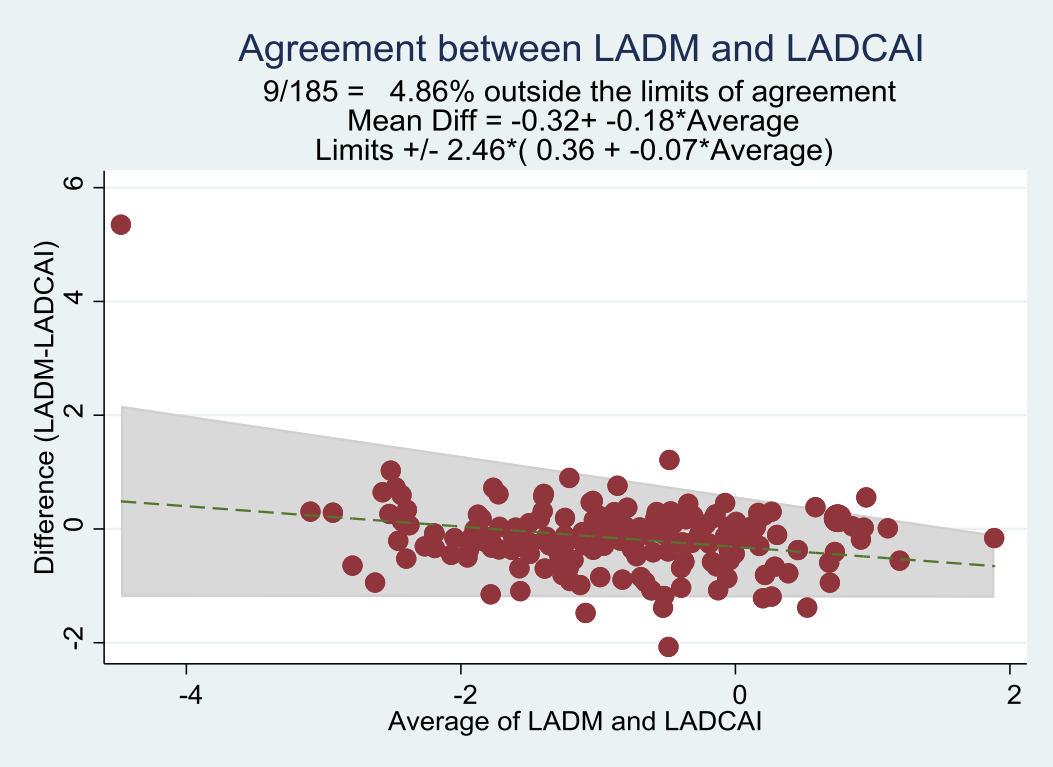
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.788[b]	.649	.876	8.428	46	46	.000
Average Measures	.881[d]	.787	.934	8.428	46	46	.000



In normal children, agreement between LADM and LADCAI

Intraclass Correlation Coefficient[a]

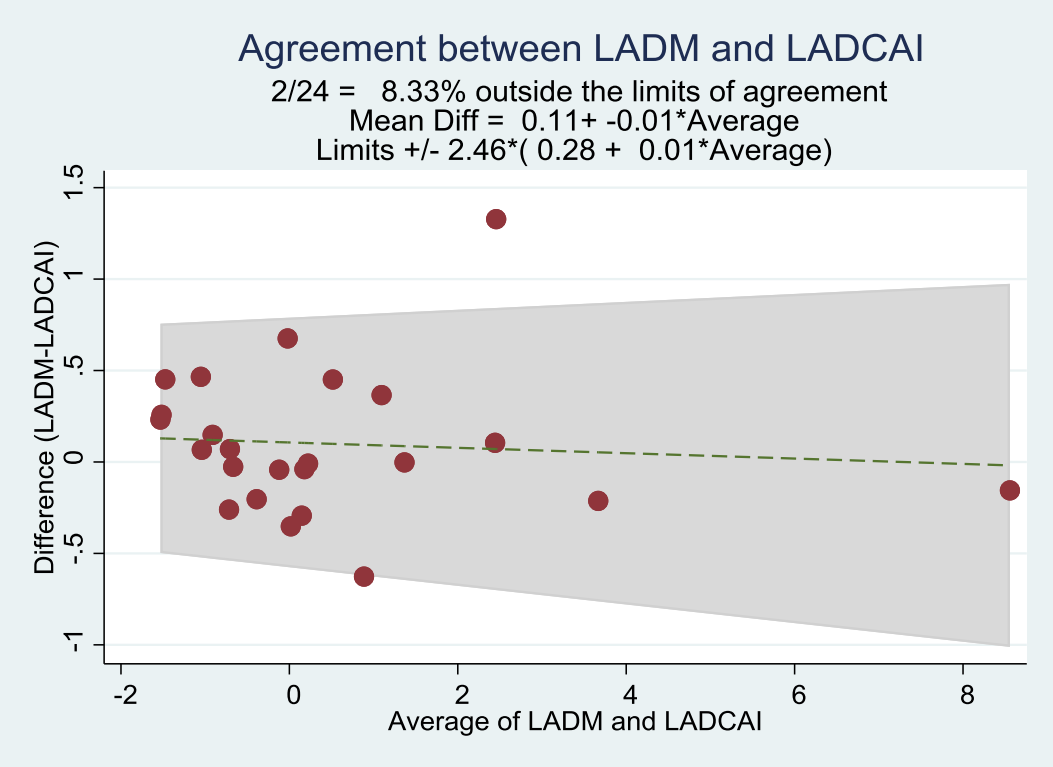
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.799[b]	.741	.846	8.963	184	184	.000
Average Measures	.888[d]	.851	.917	8.963	184	184	.000



In KD children, agreement between LADM and LADCAI

Intraclass Correlation Coefficient[a]

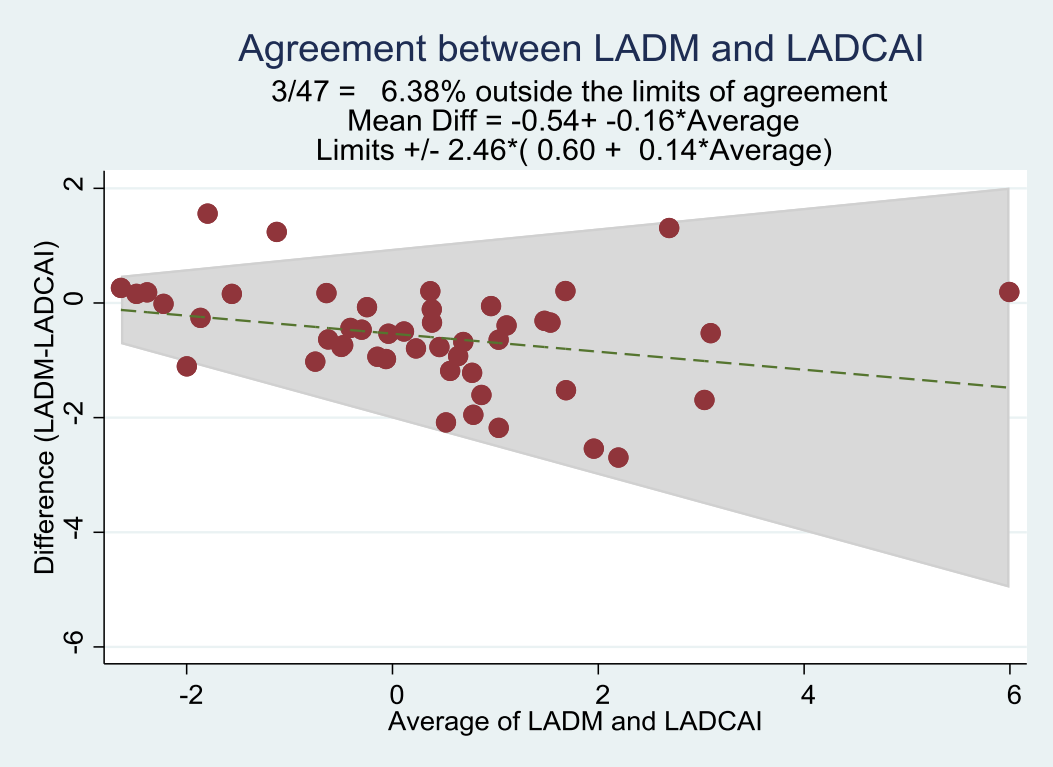
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.983[b]	.962	.993	118.190	23	23	.000
Average Measures	.992[d]	.980	.996	118.190	23	23	.000



In Misc children, agreement between LADM and LADCAI

Intraclass Correlation Coefficient[a]

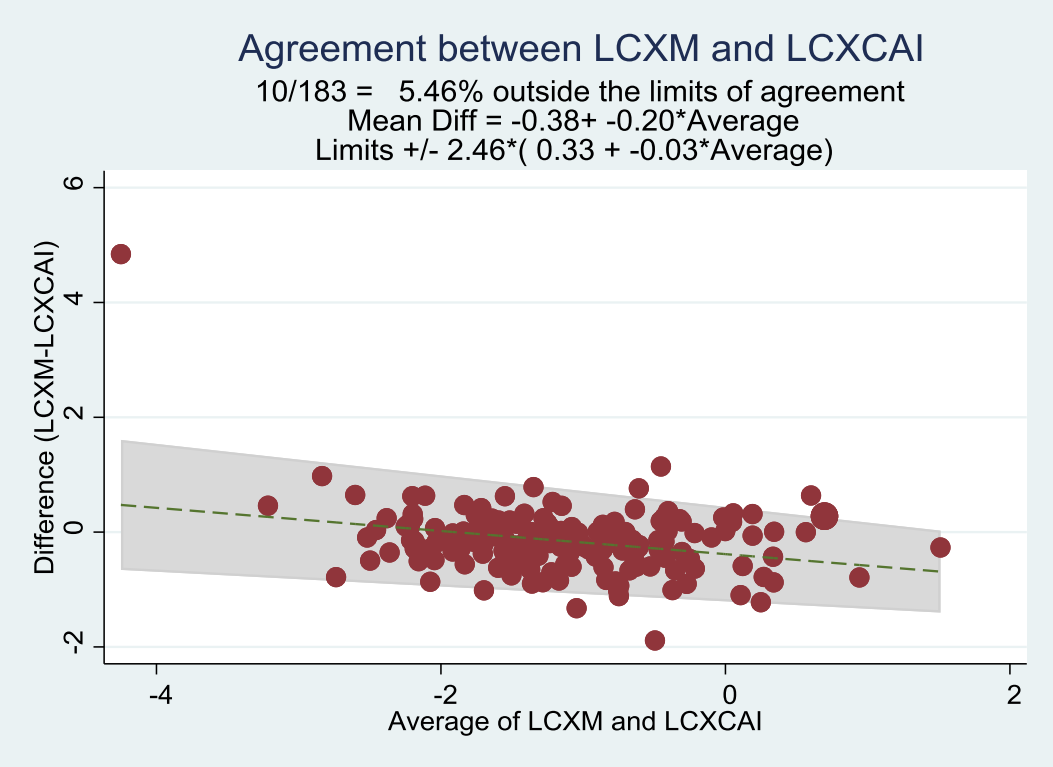
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.860[b]	.762	.920	13.311	46	46	.000
Average Measures	.925[d]	.865	.958	13.311	46	46	.000



In normal children, agreement between LCXM and LCXCAI

Intraclass Correlation Coefficient[a]

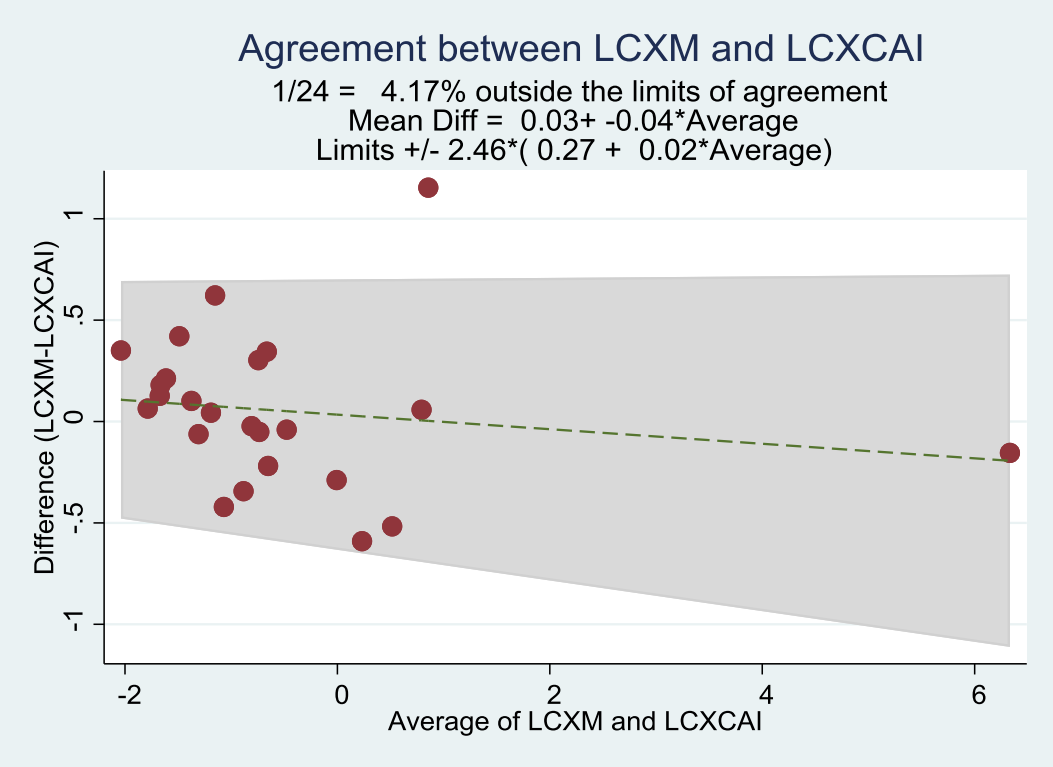
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.789[b]	.727	.838	8.469	182	182	.000
Average Measures	.882[d]	.842	.912	8.469	182	182	.000



In KD children, agreement between LCXM and LCXCAI

Intraclass Correlation Coefficient[a]

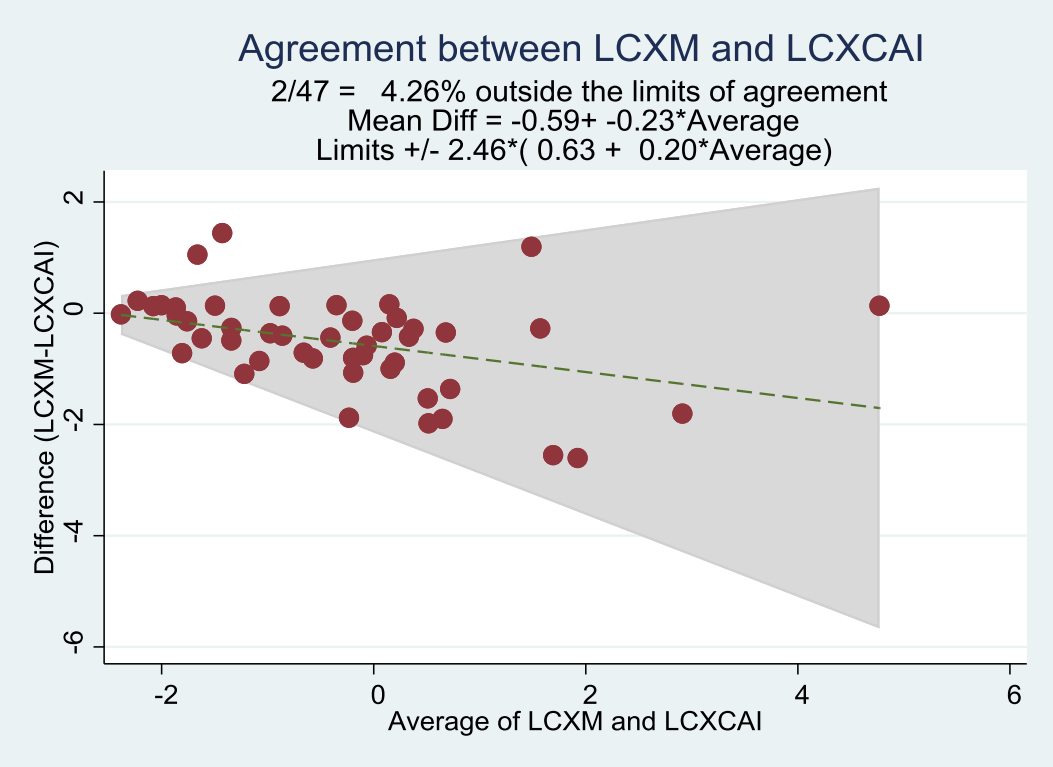
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.974[b]	.941	.989	76.452	23	23	.000
Average Measures	.987[d]	.970	.994	76.452	23	23	.000



In Misc children, agreement between LCXM and LCXCAI

Intraclass Correlation Coefficient[a]

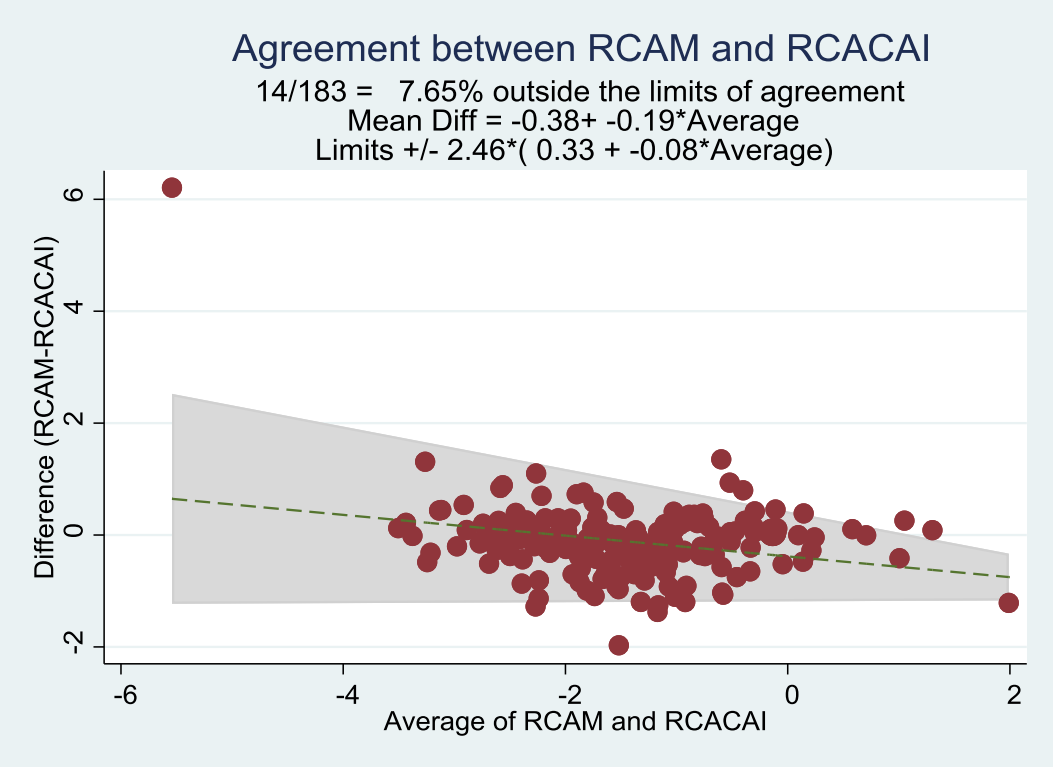
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.833[b]	.719	.904	11.001	46	46	.000
Average Measures	.909[d]	.837	.949	11.001	46	46	.000



In normal children, agreement between RCAM and RCACAI

Intraclass Correlation Coefficient[a]

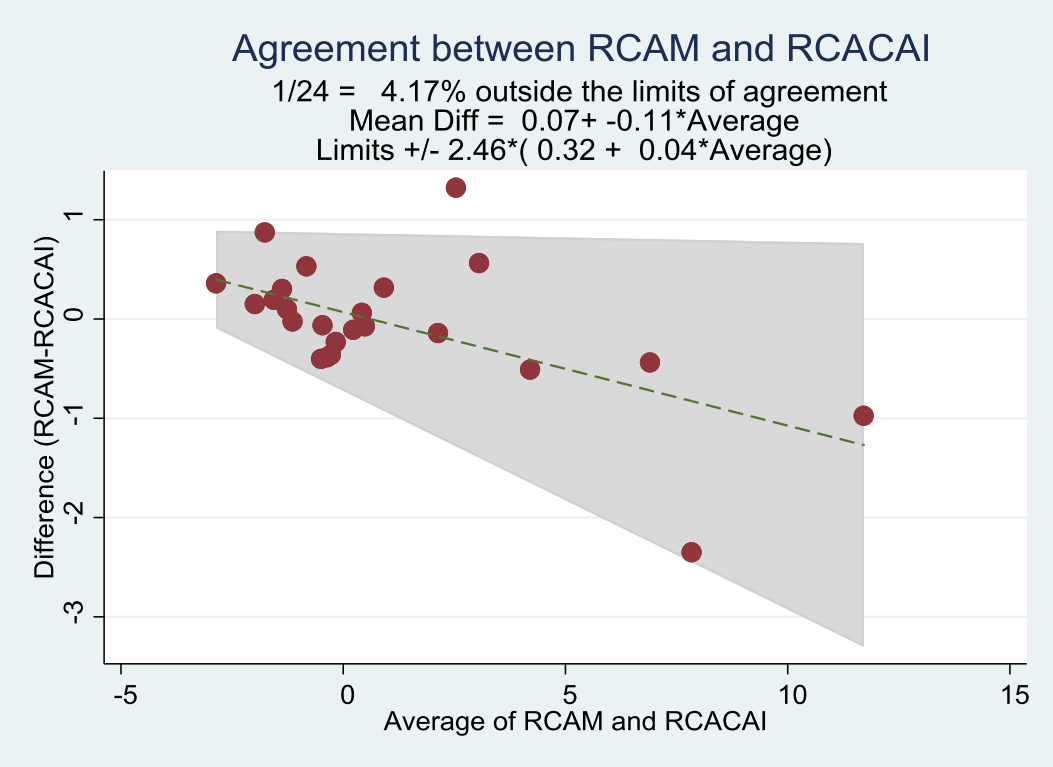
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.784[b]	.721	.834	8.250	182	182	.000
Average Measures	.879[d]	.838	.909	8.250	182	182	.000



In KD children, agreement between RCAM and RCACAI

Intraclass Correlation Coefficient[a]

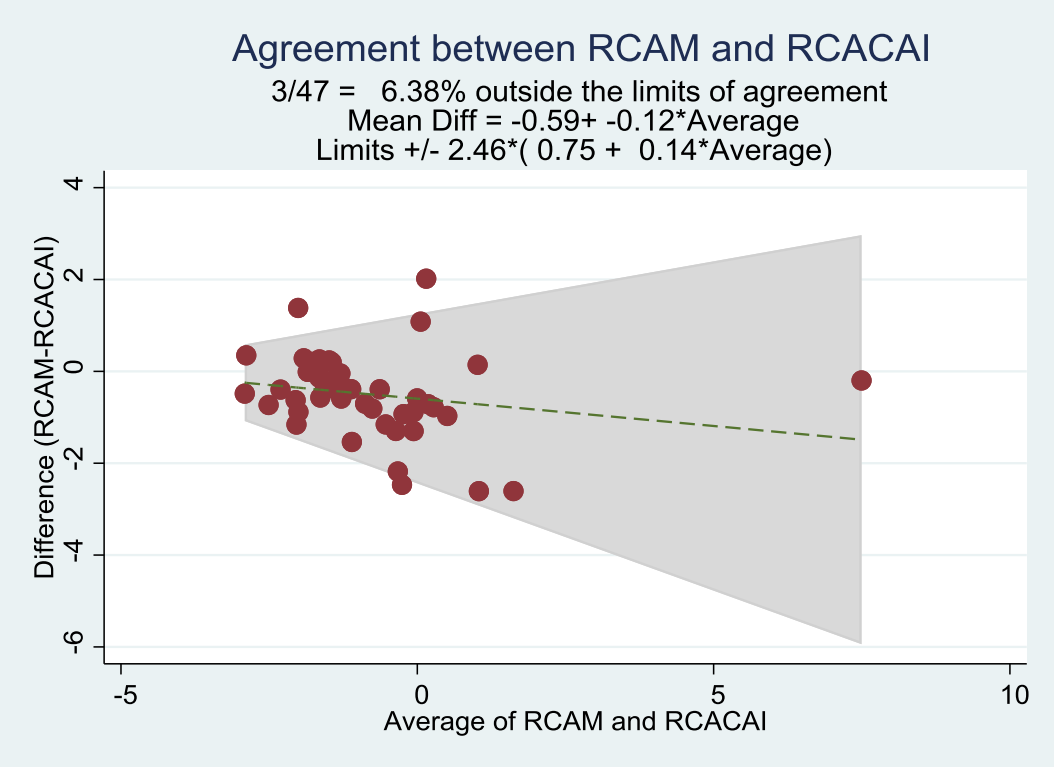
	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.981[b]	.956	.992	103.358	23	23	.000
Average Measures	.990[d]	.978	.996	103.358	23	23	.000



In Misc children, agreement between RCAM and RCACAI

Intraclass Correlation Coefficient[a]

	IntraclassCorrelation[c]	95% Confidence Interval		F Test with True Value 0			
		Lower Bound	Upper Bound	Value	df1	df2	Sig
Single Measures	.855[b]	.754	.917	12.806	46	46	.000
Average Measures	.922[d]	.860	.957	12.806	46	46	.000



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Incomplete Kawasaki Disease case series: Experience from a single center

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Introduction:

Kawasaki disease (KD) is a childhood multisystem disease of unknown aetiology characterised by vasculitis of medium sized blood vessels.It accounts for 23% of all the childhood vasculitis. KD is the second most common cause of vasculitis with Henoch Schonlein Purpura being the first with an incidence of 49%.Classical KD usually affects children in the age group of 6 months to 5 years. Incomplete KD affects children < 1 year or above 10 years. These children have persistent fever and < 4 of the 5 characteristic clinical features. Although there is no diagnostic test for KD, common laboratory features seen are raised CRP, ESR, anaemia and thrombocytosis (usually in 2nd week of illness). 2D echocardiography reveals coronary artery abnormalities. Many children have a concurrent infection, making the diagnosis of KD difficult.

In our cohort of 10 patients, five were below 1 year and five were above 10 years. In addition to persistent fever for > 5 days, children commonly had gastrointestinal symptoms such as vomiting, diarrhoea, pain abdomen followed by a polymorphous rash. Rash and pedal oedema were the common features seen in infants. 4 out of 5 of the children above 10 years presented as MIS-C with covid antibody being positive in two of them. One child below 1 year had features of MIS-C and covid antibody positivity. Two children presented in shock and one child needed inotropic support. Inflammatory markers (ESR, CRP) were raised in all the children. As KD is a diagnosis of exclusion second line investigations for fever were ordered in all children which returned normal. 2D echo showed coronary artery abnormalities suggestive of KD in nine children. All of them received intravenous immunoglobulin (IVIG) together with high dose of Aspirin. Seven out of ten children also needed steroids in view of ongoing fever. One child had to be started on Warfarin in view of persistent dilated coronaries.

Conclusion:

KD should be considered in the differential diagnosis of prolonged unexplained fever because coronary artery abnormalities occur in 15-25% of untreated KD and in approximately 5% of those who even received IVIG before 10 days of fever. Echocardiography should be performed in all cases of suspected Incomplete KD because it does help in making the diagnosis. Incomplete KD is more common in young infants (<1 year), who are at high risk to develop coronary artery abnormalities. Incomplete KD should be considered in cases where not all clinical criteria are met but coronary artery abnormalities are detected. In children >10 years, Incomplete KD is seen in association with MIS-C.

Title:

Occurrence of Kawasaki disease in temporal proximity to acute lymphoblastic leukemia – a diagnostic conundrum

Authors & Affiliation:

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Title:

Occurrence of Kawasaki disease in temporal proximity to acute lymphoblastic leukemia – a diagnostic conundrum

Background/Aims- Kawasaki disease (KD) is a common pediatric vasculitis with predominant involvement of coronary arteries. Infections have emergedas the most important triggers of KD.Neoplasms have also been reported to be associated with KD, albeit rarely and it has been postulated to trigger KD.

Methods- We report our experience of KD associated in temporal proximity with acute lymphoblastic leukemia (ALL)– the commonest childhood malignancy.Asingle-center retrospective study was conducted at Chandigarh, North India, from January 1994 to May 2021.We analysed 1078 children with KD admitted to our unit and included those who were diagnosed with ALL within 6 months of diagnosis of KD.Bone marrow examination including flow cytometry and cytogenetics was used to diagnose ALL.

Results- ALL was seen in 3 patients (0.28%). Median age at presentation was 5 years [range 4–10 years] and 2/3 patients were females. All 3 patients had incomplete KD and coronary artery abnormalities were seen in one patient.Two patients hadfever, cervical lymphadenopathy and periungual peeling at presentation and one showed perianal peeling in addition to these features. All were treated with intravenous immunoglobulin (IVIg) and aspirin with initial response. However, there was resurgence of fever in association with organomegaly and pancytopenia ranging from 1 to 3 months after diagnosis of KD. One

patient also had mediastinal mass and one had right wrist and left ankle arthritis. All patients achieved remission and no mortality was reported.

Conclusions- Higher risk of neoplasms has been reported in literature in patients with KD. However, whether the risk is higher in close temporal proximity to KD remains unknown due to paucity of reports.The question whether neoantigens on leukemic cells incite KD or unknown triggers result in both KD and ALL remains unknown. Immune dysregulation in KD has been postulated to be one of the causes for development of malignancy. Further research is needed to elucidate this association.

Cardiac Magnetic Resonance Imaging in Children with Kawasaki disease and persistent coronary artery abnormalities at a mean follow-up of 10.6 years.

Authors:

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Introduction and objectives: The most important manifestation of Kawasaki disease (KD) is development of coronary artery abnormalities (CAAs). These develop in 15-25% of patients who do not receive treatment with intravenous immunoglobulins. There is evidence to suggest that KD can also result in myocarditis that is often not clinically manifest. Although 2D-echocardiography (2DE) and CT coronary angiography have hitherto been the preferred imaging modalities , Cardiac Magnetic Resonance Imaging (CMRI) is now being increasingly recognized as a useful imaging modality for myocardial and coronary artery assessment in KD. There is, however, paucity of literature on CMRI in KD.

Methods: This prospective observational study was conducted between July 2021-September 2022 in the Paediatric Allergy and Immunology Unit, Advanced Paediatrics Centre, Department of Radiodiagnosis and Imaging and Department of Cardiology, Post Graduate Institute of Medical Education and Research, Chandigarh. Ten patients with mean interval of 10.6 years (range 9-12 years) after diagnosis of KD and with persistent CAA's underwent CMRI on 3 Tesla – Philips Ingenia platform. 2DE was also carried out the same day (Philips

Epic 7). Diagnosis of KD was based on American Heart Association guidelines (2004). All patients were on aspirin.

Results: Mean age of the study cohort was 13.8years (range 10-16 years). Of the 10 patients who underwent CMRI, 3 had ejection fraction EF <55%, 2 patients had regional wall motion abnormalities(RWMA) in septum and 1 patient hadlate gadolinium enhancement (LGE). Three patients (including 1 patient with LGE) had elevated native myocardial T1 values (>1300ms)suggestive of myocardial fibrosis. In 1 patient ,there was resolution of the CAA's that had been diagnosed previously.

Conclusion: One third of patients with KD and persistent CAAs had areas of myocardial fibrosis and LV systolic dysfunction on long term follow-up.

CMRI is a 'one stop' modality for patients with KD on long term follow-up for assessment of myocardial dysfunction. Our findings, however, need to be confirmed on a larger cohort of patients.

A RETROSPECTIVE COHORT STUDY TO IDENTIFY THE RISK FACTORS ASSOCIATED WITH RESISTANT KAWASKI DISEASEIN CHILDREN

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INTRODUCTION: Kawasaki disease (KD) is a systemic vasculitis affecting medium sized and small vessels inpaediatricpopulation.Approximately upto20% of patients with KD who remain febrile after administration of first dose of intravenousimmunoglobulin (IVIG) plus aspirin are classified as having IVIG-resistant disease. Nonresponders to initial IVIG therapy have a higher risk for coronary arterylesions (CAL), including aneurysms, compared with children who respond to first dose of IVIG.IVIG resistance was defined as fever for >36 hours after IVIG completion.

OBJECTIVE: 1)To identifythe risk factors associated with IVIG resistance inchildren which is in turn associated with higher incidence of coronary abnormalities.

2)To study the Clinical and laboratory profile of children with resistant KD.

MATERIALS AND METHODS:A retrospective cohort study was conducted in children with KD (from 1 month to 18 years of age) from January2010 to April 2018 at our single tertiary care centre. Clinical, laboratory and echocardiographic data were retrieved retrospectively.

RESULTS: Children who had persistence of fever after IVIG therapy were characterized by significantly higher values of post-IVIG white blood cell (WBC), % neutrophils and CRP whereas the IVIG responsive group showed an approximately 40%–60% reduction of WBC and CRP values.6.38% of children were resistant to initial dose of IVIG. All children resistant to IVIG had coronary artery abnormalities (coronary artery dilatation) with elevated CRP, ESR and leucocytosis ,50% of children had anaemia and hypoalbuminemia. SGOT/SGPT was normal for all six children. Demographic profile showed that all cases of resistant KD were less 2-year-oldwith 83% malepreponderance. Out of 6 cases of resistant KD who were treated with second dose of IVIG-,33% of children required other modalities of treatment in the form of Infliximab, steroids and cyclosporine. None of the children developed coronary artery aneurysm on follow up.

CONCLUSION:Scoring systems like Kobayashi, Egamihave failed to demonstrate real effectiveness to be clinically useful in predicting IVIG resistance and coronary involvement in non-Japanese populations. There is a need for development of scoring system for resistant KD children in our population and for appropriate treatment guidelines for these small group of resistant KD patients.

Retrospective Cohort Study of Clinical Profile and IVIg Resistance in Children with Incomplete Kawasaki Disease in a Tertiary Care Center

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Abstract:
Background: Diagnosis of incomplete Kawasaki disease (KD) is a clinical challenge in the absence of a specific diagnostic test. Diagnosis of KD is based mainly on typical constellation of clinical signs and symptoms. Incomplete KD is much more difficult to diagnose because they present with fewer clinical features. Hence diagnosis and treatment is often delayed in such cases. High index of suspicion is necessary to diagnose and manage the children presenting as incomplete KD.

Methods: Retrospective study of children with incomplete KD diagnosed at a tertiary care hospital from January 2010 to April 2018 was carried out. Diagnosis and treatment were based on the American Heart Association (AHA) guidelines.

Results: Fifty-four out of 94 KD cases were incomplete KD (57.4%). Most of the incomplete KD cases were in infants (48.1%). Mean duration of fever at diagnosis was 6.3 days in complete KD and 6.9 days in incomplete KD. Clinical manifestations included oral mucosal changes (79.6%), conjunctival injection (68.5%), and polymorphous exanthema (64.8%). Less common clinical manifestations were extremity changes (14.8%), cervical lymphadenopathy (33.3%), irritability (53.7%), diarrhoea (31.4%), vomiting (20.3%), and BCG scar flare up (11.1%). Coronary artery abnormalities were detected in 27 cases (50%). 49 out of 54 cases showed clinical resolution to IVIg (90.7%), 5 were resistant to the first dose of intravenous immunoglobulin, 4 responded to the 2nd dose of IVIg and one required Infliximab.

Conclusion: Incomplete KD is more often associated with CAA. Clinicians should have a high index of suspicion when fever persists for 5 days or more and is associated with any of the principal clinical manifestations. Majority of the children responded to IVIg.
Keywords: Incomplete Kawasaki disease, Intravenous immunoglobulin, Coronary artery abnormality, Infliximab

CLINICAL, LABORATORY AND ECHOCARDIOGRAPHIC PROFILE OF CHILDREN WITH KAWASAKI DISEASE AND FACTORS INFLUENCING DEVELOPMENT AND OUTCOME OF CORONARY ARTERY ABNORMALITIES

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ABSTRACT

Background- Most important step in managing Kawasaki disease (KD) is the diagnosis, as there is no single Diagnostic test. Incomplete KD with few clinical findings makes the diagnosis more difficult leading to delayed diagnosis and high risk of coronary artery abnormality (CAA).The present study reveals the clinical spectrum and outcome of KD in a tertiary center.

Methods-A retrospective cohort study was conducted in 94 children (1 month to 18 years of age) with KD from January 2010 to April 2018. Clinical, laboratory and echocardiographic data of children who were diagnosed and treated for KD according to AHA guidelines were retrieved. These children were followed up for clinical and echocardiographic outcome. Comparison was made between complete and incomplete KD in terms of clinical presentation and CAA& outcome. Primary outcome was early diagnosis, less coronary involvement and good response to treatment in complete KD.

Results-Most common clinical findings were oral mucosal changes and conjunctival injection. Most common abnormal lab parameter was elevated CRP and ESR. Incidence of incomplete KD was more than complete KD (57.4 % vs 42.6% respectively). Incidence of CAA was 42.6% (n=40). CAA was found more frequently with incomplete KD than complete KD (50% vs 32.5% respectively; *P*= 0.09). CAA was more common in children diagnosed and treated after 10 days of illness when compared to those treated before 10 days (66.6% vs 37.9% respectively; *P*=0.039). Response to one dose of IVIg was better with complete KD compared to incomplete

KD (97.5% vs 90.7% respectively). Resolution of CAA by 1 year was 100% in complete KD while it was 88.8% in incomplete KD.

Conclusion- In our study incomplete Kawasaki disease and delayed diagnosis were found to be associated with higher risk of CAA. Majority of the cases with CAA resolved during follow up and no mortality observed. Awareness among the health care providers about the illness will reduce the delay in diagnosis and also its complications.

Key words- complete Kawasaki disease, incomplete Kawasaki disease, coronary artery abnormality

MISDIAGNOSIS OF SYSTEMIC ONSET JUVENILE IDIOPATHIC ARTHRITIS AS KAWASAKI DISEASE.

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Introduction: Kawasaki disease (KD) isa diagnosis of exclusionin a febrile child lasting more than 5 days with coronary abnormalities. KD evolves over days to weeksduring which period it is diagnosed as Incomplete KD or Atypical KD. Systemic onset juvenile idiopathic arthritis (SJIA) bears a resemblance to KDwith regard to clinical and laboratory findingsin afebrile child. Here we present a 2-year-old male child diagnosed in our unit as Intravenous Immunoglobulin (IVIG) resistance KDlaterturned out to beSJIA with macrophage activation syndrome (MAS).

Case Report: 2-year-old male childwell a month back presented to local pediatric unit with a feverthat was daily,high-grade intermittent,and associated with a rash all over the body which was non-itchywith mild redness of eyes at the peak of the fever. The fever did not respond to a full course of oral antibiotics. On examination, he had hepatomegalyand cervical lymphadenopathy with irritability. Relevant investigations showed raised inflammatory markers and liver enzymes. The cerebrospinal fluid study was normal. He continued to be febrile even on broad-spectrumIV antibiotics and anti-viral given for a considerable duration for suspected meningoencephalitis. As he was still febrile with cultures and infectious workup being negative, he was referred to our unit where echocardiographyshowed dilated coronarieswith a Z score in LMCA being 5 and RCA being 4.8.As his hemoglobulin and platelets were dropping with raised ferritin and triglyceride fitting into the modified 2009 HLH criteria as well as Ravelliet al.MAS criteria,a bone marrow study was done to look for haemophagocytes which was negative. He was started on IVIG at 2 gm/kg along with pulse methylprednisolone and high-dose aspirin for severe KD.As he was afebrile for 72 hours, he was discharged with a tapering dose of steroids along with aspirin. Due to financial constraints, follow-up was delayed by almost 20 days. Fever recurred after 48 hours with daily high-grade intermittent associated with rash, redness of eyes, and irritability.Echocardiography showed adecrease

in Z score to 3 in LMCA and 2 in RCA.Laboratory findings were still consistent with HLH/MAS. He did not show BCG site reactivation, sterile pyuria, thrombocytosis, low albumin, hydrops of gall bladder,or uveitis during the entire illness. As the rash wasevanescent withintermittent febrile episodes and non-response to IVIG with non-progressive nature of coronary aneurysmin spite of continued fever, the diagnosis was revised to SJIA with MAS and started on cyclosporine and steroids. The child was afebrile after a week and playful with a drop in ferritin (11343) and liver enzymes.

	At the onset	During MAS	Before Cyclosporine
Hb in gm/dl	10.8	9.0	8.6
TLC per cumm (N/L)	39700 (58/32)	14300 (46/50)	5490 (11/78)
TPC per cumm	165000	143000	330000
ESR (mm/hr)/ CRP (mg/dl)	37/-	46/86	2/ 130
Total Protein/Albumin (gm/dl)		6.7/3.5	6.7/3.8
SGOT/SGPT IU/ml		200/70	249/102
LDH/ Triglyceride in mg/dl		-/555	1708/433
Ferritin in ng/ml		1650	100000
Il-6 in pg/ml and d-dimer µg/dl		64.8 / 1.5	-/3500

Conclusion: Coronary artery abnormalities are reported in various vasculitis. SJIA should be kept as differential in a child diagnosed with KD not responding to IVIG and manifesting as MAS. Surrogate clinical and laboratory features help us to differentiate both diseases. IVIG affordability in a resource constraint country like ours makes it difficult for a second dose.Stringent follow-up of such cases for future complications and treatment with aggressive immunomodulation as they carry a bad prognosis.

Intestinal obstruction with febrile illness - "Keep your eyes peeled for KD"

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Background:

Kawasaki disease is a vasculitis of unknown origin of medium caliber blood vessels, especially involving coronary arteries. Gastrointestinal symptoms are commonly observed in KD however intestinal pseudo-obstruction is uncommon. We hereby present one such unusual case of Kawasaki disease.

Case report: 16 month old girl presented with fever, bilious vomiting, loose stools. On examination, she was dehydrated and had abdominal distension. She also appeared toxic and had an erythematous rash over vulval area. Erect x-ray abdomen and Ultrasound abdomen showed signs of small bowel obstruction. She was started on conservative management, kept NPO, and treated with IV fluids, IV antibiotics and other symptomatic management. On Day 2 of admission, child had persisting fever spikes and episodes of bilious vomiting. Repeat blood investigations done showed a marked elevation in inflammatory markers. CECT Abdomen was done and confirmed the small bowel dilatation. At this point, features of conjunctival congestion, hyperemia of BCG scar were seen and she was diagnosed as atypical Kawasaki disease. She was treated with IVIg and low dose IV methylprednisolone which led to marked improvement, both in terms of defervescence and disappearance of signs of intestinal obstruction.

Conclusion :

Intestinal obstruction is an uncommon (2.3 %) but important manifestation of Kawasaki disease though vomiting and diarrhea are frequently reported symptoms. Hence, a high suspicion for small bowel obstruction due to mesenteric vasculitis secondary to KD in children with febrile illness is essential not only to reduce diagnostic and therapeutic delays leading to risk of development of coronary complications in KD but also to prevent unnecessary surgical intervention which may lead to high mortality.

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INFANTILE KAWASAKI DISEASE: A DISTINCT ENTITY?

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BACKGROUND:

Kawasaki Disease (KD), an acute febrile medium vessel vasculitis is predominantly seen in children less than five years. Due to the absence of any specific biomarker diagnosis is clinical. Children less than one year of age mostly present with fewer signs and symptoms, may even be masked by other non-specific manifestations, hence the diagnosis of KD often gets delayed with added delayed referrals. Delayed diagnosis and hence late initiation of intravenous immunoglobulin (IVIG) increases the incidence of coronary artery abnormalities(CAA). KD in less than 3months is seldom reported and there is paucity of data on infantile KD.

METHODS

This is a retrospective, analytical study on data of children less than or equal one year of age diagnosed as KD defined as per the AHA guidelines, at Institute of Child Health Kolkata from January 2018 to September 2022.

RESULTS

136patients were diagnosed as KD from January 2018 to September 2022,29(21%) being infants. Median age of the infants was 180 days (IQR 160), the youngest being 6 weeks old. 15 were less 6 months and 14 more than 6months. 15 were males and 14 were females. Majority ie. 76% (n=22) of infants presented as incomplete KD;73% in less than 6 months and 85% in more than 6months. The mean duration of illness at admission was 8 days and mean duration of illness when first dose of intravenous immunoglobulin (IVIG) was administered was 11 days. Fever of more than 5 days was present in 86%, mucositis was seen 62%, non-purulent conjunctivitis in 62%, rashes (ranging from a generalised maculopapular rash, erythema multiforme like rash to erythroderma) in 55%,unilateral lymphadenopathy in 45%, and dorsal oedema of extremities in 45%. Other symptoms were also seen such as periungualdesquamation (34%), BCG site reactivation (28%), orange brown chromonychia in (31%). 28% had hepatomegaly and 7% had sterile pyuria. Apart from fever, mucositis and conjunctivitis was common in both the age subgroups. One baby had convulsions and one had persistent tachycardia.

On admission, the meanhemoglobin was 8.86gm/dl(it was found to be lower in babies less than 6 months), TLC 21522/cmm, ANC 9920, platelets 653000/cmm, CRP 137.83 mg/L.

IVIG was given to all. 10% (n=3) were resistant to IVIG and were given infliximab (IFX).

12 (41.4%) babies had coronary artery aneurysms (CAA) at diagnosis. 8 (57.14%) had CAAs in more than 6 months group vs 4 (26.66%) in those less than 6 months. Small CAA's (2.5 - 5z score) were seen in 17% (n=5), mediumsized (5 - 10z score) in 17% (n=5) and giant CAA (>10z score) in 6% (n=2)One 2 months old baby with late diagnosis after 3 weeks had multiple giant aneurysms with large thrombus in LAD, was IVIG resistant and needed thrombolysis. 4/5 of patients with moderate CAA at diagnosis received additional IFX and 1 received steroids.

CONCLUSION

Compared to our overall data on KD, those presenting in infancy had more incomplete presentations (76% vs 20%), with consequent late diagnosis (day 11 vs day 7) and higher incidence of CAAs (41.4% vs. 22%). However, majority of them were IVIG responsive (90% vs 80%).

INFLIXIMAB IN IVIG RESISTANT KAWASAKI DISEASE: EXPERIENCE FROM A TERTIARY CARE CENTER IN EASTERN INDIA

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Institute of Child Health, Kolkata.

BACKGROUND:

10-20% of Kawasaki Disease (KD) patients do not respond to a single dose intravenous immunoglobulin (IVIG), warranting further management.

METHODOLOGY:

This is a retrospective observational **s**tudy conducted at Institute of Child Health, Kolkata from January 2016 to September 2022. KD patients with persistent or recrudescent fever after 36 hours of IVIG infusion were enrolled. These children received Infliximab (IFX) 5mg/kg single dose. Data wasanalysed for defervescence of fever in hours and normalization of inflammatory markers (CRP at 48 hours after IFX infusion).

RESULTS:

210 children were diagnosed with KD during the study period, 31 (14.76%) were IVIG resistant and received IFX.29 received infliximab and 2received the biosimilar of infliximab.The median age was 23 months (IQR 27).10 (32.26%) were ≤12 months, 20 (64.52%) were >12mo but <60mo and 1 (3.22%) was more than 60 mo. 23 were males and 8 were females (male:female ratio 2.86:1).Mean duration of illness at presentation was 6.73±1.70 days. Mean duration of fever before administering IVIG was 7.48.±2.57 days.The median time for administration of IFX was 3 days after completion of IVIG administration.

7 of these 31 children had coronary artery abnormality (CAA) at presentation.

Following IFX administration, 29 became afebrile within 24 hours of IFX administration, remaining 2 within 48hours. The response to IFX in terms of defervescence and normalization of CRP was statistically significant (Table 1). There were no adverse reactions to IFX.

Table 1 : Response to Infliximab in IVIG resistant KD patients

	Before IFX	**After IFX**	**P value**[#]
Fever (days)	14.8 ± 5.6	1.9 ± 0.83	<0.0001
Mean CRP (N:<5 mg/ l)	74.08 *	3.96**	<0.0001

* post IVIg
** 48 hours post IFX

[#] P value < 0.05 is considered as significant in this study

CONCLUSION:

Our study cohortshowed good response to IFX in IVIG resistant cases with rapid resolution of fever and normalization of raised inflammatory markers.

Is it all Kawasaki disease? A case report

Manjari Agarwal, Sujata Sawhney

Sir Ganga Ram Hospital

16 mo. old girl was referred with a diagnosis of atypical and resistant Kawasaki disease.

Child was unwell from 13 months of age when she had multiple episodes of vomiting and poor oral intake. These lasted for 4 days and child required intravenous fluids as well.Had one more episode of vomitings lasting for 5 days with poor oral intake and irritability. Few bouts of vomitings were bilious.After a few days again had multiple episodes of vomiting,fever Developed right sided facial asymmetry. MRI brain was suggestive of acute hemorrhagic infarct of basal ganglia.MR angiography was normal.Child had high grade fever and elevated acute phase reactants.

In view of fever with elevated ESR,CRP and platelets and hemorrhagic infarct, child was treated a sincomplete, atypical Kawasaki disease and received IVIG and steroids and was discharged.There was mild dilatation of coronary artery, LMCA z score +2. There were no clinical features suggestive f Kawasaki disease. Her COVID antibodies were elevated.

Steroids were gradually tapered and stopped. Again she had high grade fever with elevated CRP. She was treated as resistant Kawasaki disease and received Infliximab this time. Child did not have any fever for 10 days and again had fever low to moderate grade with irritability.

She is the first child born of non consanguinous marriage. Her antenatal, birth and first year of life were unremarkable. She had received primary immunization and was a well grown child with weight and length at 50^{th} centile.

She was referred at our centre after 8 weeks of illness and on detailed history, it was determined that the periods of fever were interspersed with normal afebrile period.

On examination she had remarkable livedo racemose.

CT angiography of abdomen was normal. 2 D echo was normal.

ADA2 enzyme assay was sent which was 0% of total ADA activity.

Mutation analysis was sent by previous team and was suggestive of homozygous mutation in ADA2 gene on exon 2 variant c.139G>A (p.Gly47Arg)

She was thus given a revised diagnosis of deficiency of ADA2.

She was commenced on Etanercept and is currently doing well.

Conclusion: Stroke in a young child with livedo needs careful evaluation. Livedo was present in this child from before these episodes.

A high index of suspicion is required for a diagnosis of DADA2 which may have varied presentations.

Giant coronary Artery Aneurysms: A 10 year long term follow up retrospective study

Manjari Agarwal, Sujata Sawhney

Background: Giant coronary artery aneurysm is a dreaded complication of Kawasaki disease due to relentless inflammation. Long term sequelae of these are not yet well understood.

Aims: We undertook this retrospective review to evaluate long term sequelae of children with giant coronary artery aneurysms

Methods: All children with giant coronary artery lesions were included for review.

Data was collected on predesigned proformas. Treatment details and serial echocardiographic findings were noted.

Results: 211 children with kawasaki disease were seen on follow up visits in our unit from October 2012 to September 2022. 12 children were identified with giant coronary artery aneurysms.

There demographic and clinical details are tabulated in table 1.

There was no mortality in this cohort.One child has presented with thrombus.Delay in diagnosis of more than 15 days was present in most. Atorvastatin was given to all.

On last follow up, 6 children had normal coronary artery dimensions. Three children underwent CT coronaries and demonstrated resolution and normalisation of the coronary aretries

Conclusion: Long term follow up into adulthood is needed for these children even after resolution of aneurysms. Modification of lifestyle and avoidance of risk factors for increased cardiovascular morbidity such as obesity, hyperlipidemia and hypertension require strict monitoring.

Table 1: Clinical profile of children with Giant CAAs

N=11/211 Boys: Girls	5.2% 10:1
Median age at presentation(years)	0.75 years (IQR 0.34-4.9)
Complete : Incomplete presentation	2:9
Median delay to diagnosis	16 days(IQR 6-31)
IVIG 1st dose	11
IVIG 2nd dose	1
Steroids	11
LMW heparin	11
Infliximab	2

Ciclosporin	2

Infantile versus Non-infantile Kawasaki Disease – A comparative analysis from a tertiary care centre in South India

Authors: Neha Singh[1], Rachna Shanbhag Mohite[1], Ramya S[2], Jeeson Unni[3], Suresh Kumar[4], Sujatha Thyagarajan[5], Jyothi Raghuram[6], Rajappan Pillai[3], Gladys Cyril[3], George Paul[3], Sathish Kumar[4], Manjula Anand[4], Vinitha Anirudhan[4], Sangeetha Budur[5], Sindhu M V[5], Shrinivas Murthy[6], Lathiesh K Kambam[6], Arun Kumar[6],Karthik Arigela[2], Syed M Naushad[2], Chetan Ginigeri[2], Sagar Bhattad[1]

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Introduction
Kawasaki disease(KD) is an acute multisystem inflammatory disease involving medium sized blood vessels (vasculitis) that most commonly affects infants and young children.

Objectives:
To study and compare the clinical profile of patients diagnosed with infantileand non-infantile KD at a tertiary care centre in Bangalore, India.

Methods:
A retrospective review of clinical records was performed and patients with KD diagnosed during the study period (Feb 2017 to October 2022) were included. The clinical profile, treatment and complications were compared between the infantile and non-infantile KD group. A detailed analysis was performed to understand the similarities and differences in the presentation, response to treatment and outcomes of patients.

Results:
A total of 64 patients with KD were included, out of which 24 had infantile KD. Males were more commonly affected than females (Table 1) in both groups. The mucosal changes, cervical lymphadenopathy, non-exudative conjunctivitis and rashes was more frequent in infantile group while cervical lymphadenopathy and extremity changes was more common in non-infantile group. In systemic manifestations, diarrhoea and irritability was more frequent in infantile group. Fifty percent infantile KD patients developed coronary aneurysms, while 35% non-infantile KD patients developed aneurysms. The haematological parameters showed increased leucocyte count in infantile KD as compared to non-KD group. Fifty percent infantile KD patients had IVIg resistant KD, while 30% patients in the non-infantile KD group required second line immunomodulation.

Conclusion:

This study highlights the clinical similarities and differences among patients with infantile and non-infantile KD. Coronary aneurysms and IVIg resistance was more frequent in infantile KD.

Table 1. Clinical characteristics and treatment of Infantile and non-infantile KD patients

	Infantile Kawasaki Disease (n=24)	**Non infantile Kawasaki Disease(n=40)**
Age(in months)	7.67	56.1
Male	18(75%)	27(67.5%)
Female	6(25%)	13(32.5%)
Mucosal changes	20(83.3%)	22(55%)
Non exudative conjunctivitis	18(75%)	18(45%)
Cervical lymphadenopathy	2(8.3%)	13(32.5%)
Rashes	23(95.8%)	22(55%)
Extremity changes	7(29.2%)	15(37.5%)
Perianal peeling	5(20.8%)	5(12.5%)
Systemic involvement		
Shock	1(4.2%)	2(5%)
Diarrhoea	6(25%)	2(5%)
Respiratory symptoms	2(8.3%)	4(10%)
Seizures	2(8.3%)	1(2.5%)
Irritability	13(54.2%)	11(27.5%)
Cardiac involvement		
Aneurysms*	12(50%)	14(35%)
LAD	9(37.5%)	12(30%)
Small	6	7
Moderate	3	2
Large	-	3
LcX	2(8.3%)	1(2.5%)
Small	1	1
Moderate	1	-
LMCA	5(20.8%)	5(12.5%)
Small	-	2
Moderate	3	1
Large	2	2
RCA	5(20.8%)	2(5%)
Small	3	1
Moderate	2	1
Pericardial effusion	1(4.2%)	2(5%)
Aortic root dilatation	2(8.3%)	3(7.5%)

Valvular regurgitation	3(12.5%)	3(7.5%)
Myocardial infarction	-	1(2.5%)
Average duration of fever(days)	9.6	9.5
Mean duration for defervescence after IVIg(days)	1.2	1.2
Mean duration of hospital stay(days)	6.2	4.1
Haematological Parameters		
Haemoglobin(g/dl)	9.6	10.8
Total counts(cells/cu mm)	16152	13533
Platelet counts(/cu mm)	680,000	544,000
CRP(mg/l)	73.8	75.3
ESR(mm/hr)	68.4	66.3
Treatment		
IVIG	23(95.8%)	39(97.5%)
IVIG Resistance	12(50%)	12(30%)
Methylprednisolone	12(50%)	12(30%)
Infliximab	4(16.7%)	6(15%)
Supportive therapy		
Aspirin	24(100%)	39(97.5%)
Dual antiplatelet agents	1(4.2%)	2(5%)
Warfarin/LMW Heparin	2(8.3%)	2(5%)

LAD - left anterior descending artery, LMCA - left main coronary artery, LcX - left circumflex artery, RCA – right coronary artery

Acquired phimosis in an infant with IVIG-resistant Kawasaki Disease: A rare occurrence

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Introduction:

Urethritis in Kawasaki disease (KD) is well known to cause sterile pyuria and is secondary to non-specific vasculitis of urethra. We hereby report a case of a male infant with Kawasaki disease who had urethritis during the hospital stay, and presented a week later with phimosis.

Case report:

A 7-month-old boy, born to Indian parentage, presented with high grade feverfor 20 days. The fever was associated with redness of lips and strawberry tongue on fifth day of illness, with mild extremity swelling, followed by peeling of skin in perianal regiona week later(Figure 1). He was diagnosed with KD and treated with IVIg(2 g/kg) elsewhere, however fever did not subside. On examination, he had severe pallor, and desquamative changes in the perineal regionincluding the preputial skin. He was hemodynamically stable, and the systemic examination was normal, however was noted to have remarkable irritability. The baseline investigations done showed anemia (5.4 g/dL) and thrombocytosis (10,19,000/ cu mm) The inflammatory markers were elevated (CRP 271 mg/L). The ferritin was 204 ng/ml and pro BNP was 269 pg/ml. The echocardiogram showed small aneurysms in left anterior descending (LAD) (2.8 mm, Z Score 3.8) and left main coronary artery (LMCA)(2.7 mm, Z score 4.9). He was treated with intravenous Methyl Prednisolone (IV MP)(2 mg/kg) and IV Infliximab (10 mg/kg) He responded promptly, irritability decreased and he became afebrile. CRP showed decreasing trend (271 - 107 mg/L). IV MP was given for 3 days and later switched to oral steroids. Aspirin was given at anti-platelet dosage. Echocardiography done prior to discharge showed some improvement in the coronary arteries with mild aneurysm of LAD (LAD 2.0 mm Z Score 2.77).

He presented a week later with transient ballooning of penile shaft while micturating. He also had developed dribbling and poor urine stream after discharge. On examination, he had phimosis. (Figure 2) He was referred to the pediatric surgery services and was planned for circumcision.

Conclusion:

We describe a rare and serious complication of KD, which would have long term implications if missed.

Kawasaki disease versus Multi-system Inflammatory Syndrome in Children (MIS-C) – A comparative analysis from a tertiary care centre in South India

Authors: Neha Singh[1], Rachna Shanbhag Mohite[1], Ramya S[1], Jeeson Unni[2], Suresh Kumar[3], Sujatha Thyagarajan[4], Jyothi Raghuram[5], Rajappan Pillai[2], Gladys Cyril[2], George Paul[2], Sathish Kumar[3], Manjula Anand[3], Vinitha Anirudhan[3], Sangeetha Budur[4], Sindhu M V[4], Shrinivas Murthy[5], Lathiesh K Kambam[5], Arun Kumar[5],Karthik Arigela[1], Syed M Naushad[1], Chetan Ginigeri[1], Sagar Bhattad[1]

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Introduction

Multi System Inflammatory syndrome in Children (MIS-C) is now one of the well-recognised complications of COVID-19 infection seen mostly in the paediatric patients. MIS-C has clinical features similar to those found in Kawasaki disease (KD), macrophage activating syndrome and toxic shock syndrome.

Objectives:

To study and compare the clinical profile of patients diagnosed with MIS-C and KDat a tertiary care centre in Bangalore, India.

Methods:

A retrospective review of clinical records was performed and patients withKDdiagnosed during the studyperiod (Feb 2017 to October 2022) were included. The clinical profile, treatment details and short-term outcomes were compared with a large cohort of patients with MIS-C diagnosed during the COVID pandemic (Aug 2020 to March 2022). A detailed analysis was performed to understand the similarities and differences in the presentation, response to treatment and outcomes of patients.

Results:

A total of 64 patients with KD and 81 patients with MIS-C were included in the study. The mean age of presentation in children with KD (3 years) was younger as compared to patients with MIS-C (6.8yrs). Males were more commonly affected than females (Table 1) in both groups. The mean duration of fever prior to presentation was 9.5 days in KD compared to 4.6 days in MIS-C. Conjunctival congestion, rashes and lymphadenopathy were common symptoms in both groups. Gastrointestinal symptoms (abdominal pain, loose motions) (53% vs 10.9%), cardiovascular shock (26.9% vs 4.6%) and myocardial dysfunction (54.3% vs 3.1%) was significantly higher in patients with MIS-C. On the other hand, 40.6% (26/64) KD patients had coronary artery aneurysms (CAA) involving single (19/26) or multiple coronary arteries (7/26) as compared to only 9.8% patients with MIS-C. Lymphopenia (29.6%) was characteristic of patients with MIS-C (29.6%).Most children (66.6%) with MIS-C were treated with a combination of IVIG and steroids whereas IVIG (96.8%) was the first line of treatment

in patients with KD and steroids (37.5%) were added as per standard indication. Use of biologics was higher in KD patients (Infliximab- 15.6%) versus 3.7% (Anakinra, Infliximab) in MIS-C. Five children with MIS-C responded to conservative management and did not receive any immunomodulation. Macrophage activation syndrome was noted in a single MIS-C patient. All MIS-C patients with significant cardiac dysfunction(54.3%) received short term (6 weeks) anti-coagulation with low molecular weight heparin as compared to 9.3% KD patients who remained on long-term anti-coagulation. All children recovered and follow-up ECHO at 6 weeks was normal in all patients with MIS-C whereas 23.4%KD patients continued to have CAA or valvular involvement on ECHO.

Conclusion:

This study highlights the clinical similarities and differences among patients with KD and MIS-C. Though hemodynamic instability and cardiac dysfunction were prominent findings in MIS-C, all patients had rapid-resolution followingimmunomodulation therapy. However, long term morbidity was seen in significant number of patients with KD.

Table 1: Demographic & clinical features of MIS-C and KD patients

Clinical features	**MIS-C** **N=81**	**Kawasaki disease** **N=64**
Age in years (mean)	6.8yrs	3yrs
Male: Female	2.5:1	2.3:1
Mean duration of fever (days)	4.6	9.5
Conjunctival congestion	44 (54.3%)	36 (56.2%)
Skin rash	44 (54.3%)	45 (70.3%)
Lymphadenopathy	20 (24.6%)	15 (23.4%)
GI symptoms	43(53%)	7 (10.9%)
Neurological symptoms	8 (9.8%)	3 (4.6%)
Shock	24 (29.6%)	3 (4.6%)
Others	-	Perianal skin peeling – 10 (15.6%)
Platelet count (/mm^3)	1,73,865	6,10,700
CRP (mg/dl)	151	74.6
ESR (mm/hr)	47	71
LV dysfunction	46 (54.3%)	2 (3.1%)
Coronary artery aneurysms	8 (9.8%)	26 (40.6%)
Conservative management	5 (6.2%)	0

IVIG	11 (15.3%)	62 (96.8%)
IVIG +Steroids	54 (66.6%)	23 (35.9%)
Steroids	63(77.7%)	24 (37.5%)
Infliximab	0	10 (15.6%)
Anakinra, Tocilizumab	3 (3.7%)	0
Death	0	0

Title: Neurological manifestations of Kawasaki disease - Our experience at tertiary care center from North-Western India

Author: Pallavi L Nadig, Rakesh Kumar Pilania, Ankur Kumar Jindal, Suprit Basu, Reva Tyagi, Deepti Suri, Amit Rawat, Surjit Singh

Introduction:

Kawasaki disease (KD) is a medium vessel vasculitis, presenting typically inchildren below 5 with characteristic features such as polymorphous rash, extremity changes,mucosal changes and conjunctivitis and cervical lymphadenopathy. Neurological features likecerebrospinal fluid pleocytosis, seizures, facial nerve palsy, paralysis of the extremities havebeen described. We herein report our experience on neurological manifestations in our cohortof KD.

Patients and methods: All children with KD from Chandigarh, who presented to Pediatric AllergyImmunology unit, Postgraduate Institute of Medical Education and Research, Chandigarh, atertiary care center in north India from January 1994 - September 2022 were analyzed.

Results: Neurological manifestations were noted in 20 of 1187 patients (1.68%). Mostcommon manifestation was generalized seizures which was seen in 9 patients. Others includedencephalopathy (n=6), focal seizures (n=1), headache (n=4), transient visual loss (n=1),features of raised intracranial pressure (headache, extensor plantar, neck rigidity, papilledema)(n=2), and sensory neural hearing loss (n=1). Neuroimaging was carried out in 12 patients withvarious findings including transient hydrocephalus (resolved on follow-up imaging) (n=1),cerebral venous thrombosis (n=2), micro bleed (n=1), hypoxic injury (n=1), ring enhancinglesion (n=1), and normal study in rest (n=6) of the patients. Lumbar puncture

and cerebrospinalfluid (CSF) examination was done in 11 patients; 3 had SF pleocytosis, in rest 8 patients CSFstudy were normal. Concomitant gastrointestinal manifestations were seen in 6 patients,pulmonary in 5, KD shock syndrome in 2, macrophage activation syndrome in 1, scrotalgangrene in 1, and arthritis in 1. Coronary artery abnormalities were seen in 6 patients(coronary artery aneurysm in 2, coronary artery dilatations in 4 patients).

Conclusion: Neurological manifestations, though rare, are important cause of confusion in diagnosis of KD due to presentation which is indifferentiable to other causes of acute encephalopathies.

Title: **Use ofInfliximab in90 children with Kawasaki Disease: Experience from a tertiary care centre in North-West India**

Authors: Prabal Barman[1], Ankur Kumar Jindal[1], Rakesh Kumar Pilania[1], Pandiarajan Vignesh[1], Reva Tyagi[1], Archan Sil[1], Suprit Basu[1], Saniya Sharma[1], Manpreet Dhaliwal[1], Deepti Suri[1], Amit Rawat[1], Manphool Singhal[2], Surjit Singh[1]

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Abstract:

Background: Kawasaki disease (KD) is a medium vessel vasculitis of agnogenicorigin. Although intravenous immunoglobulin (IVIg) is the standard of care, however, approximately 20% of patients remain refractory to conventional therapy. In addition, this subset of patients with KD has a higher propensity to develop coronary artery abnormalities (CAA). It has been reported that infliximab can produce rapid defervescence and reduce CAA in IVIg-resistant and severe forms of KD. We report the role of infliximab in 90 children with KD.

Aims: To reportour experience withinfliximabin KD from a tertiary care centre in North-West India.

Methods:A review ofmedical records of all patients who were diagnosed to have KD during the period January 1994 - September 2022 in Pediatric Allergy Immunology Unit, Advanced Pediatrics Centre, Postgraduate Institute of Medical Education and Research,

Chandigarh, India was done.Case records of children with KD who hadreceived infliximab were analysed in detail.

Results: Of the 1187 patients with KD, 90 patients (63 boys) receivedinfliximab.Median age at diagnosis was 2 years (range 0.3-17 years). Indications for using infliximab were: IVIg resistance (13/90); presence of CAA (3/90); both IVIg resistance and CAA (65/90); macrophage activation syndrome/myocarditis (5/90; 1 also had CAA); retinal vasculitis (1/90); arthritis (1/90); and economic reasons (2/90). Amongst the patients who had CAA, 30 (33%), 9 (10%) and 30 (33%) had mild, moderate and giant aneurysms respectively. Twenty-nineand 14 patients also received steroid, and steroid with cyclosporin respectively because of persistence of inflammatory parameters. Dimensions of giant CAA normalised, and reduced in size, in 8 and 11 patients, after a median duration of 315 days (range 50-480 days), and 1095 days (range 180-4380 days) respectively. Moderate and mild CAA normalised in 5 and 26 patients after a median duration of 730 days (range 400-2190 days) and 27 days (range 2-520 days) respectively. None of the patients had any adverse event during transfusion nor did any patient have any significant infection over a cumulative follow-up of 5851 patient-months.

Conclusions: Infliximab may be a useful alternative for treatment augmentation in severe KD and also for management of resistant KD.

Characterization of endothelial dysfunction markers -circulating endothelial cells and endothelial progenitor cells by flow cytometry in patients with Kawasaki disease from North India

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lntroduction:

Kawasaki disease (KD) is a systemic, medium vessel vasculitis. Cardiovascular complications are major concerns associated with KD. The endothelium plays an important role in maintaining vascular homeostasis. Any kind of stress or injury can lead to endothelial dysfunction that is an important pathogenic event in cardiovascular diseases. Circulating endothelial cells (CECs) are associated with vascular injury and circulating endothelial progenitor cells (EPCs) are capable of vasculogenesis and have association with cardiovascular risk factors.

Methods:

The present study is a single centre, prospective study in North India to characterize CECs and EPCs in patients with Kawasaki disease. Study includes KD patients (diagnosed as per American Heart Association **(AHA) criteria 2004**) at different time intervals with and without coronary artery aneurysms (CAA) (**Z score as per Montreal (JASE 2011)**. KD patients with CAA were further classified into transient and persistent aneurysms as per **AHA 2017**.

Four groups were enrolled-

Group 1: Diagnosed prior >6 months-1.5 years **(N=19)**

Group 2: Diagnosed >1.5 - 3 years **(N=24)**

Group 3: Diagnosed > 3 - 4.5 years **(N=22)**

Group 4: Age and sex matched healthy controls **(N=20)**

Maximum patients (56%) were <5 years of age while 12% were > 10 years of age. Median age was 4 (range: 0.2-13) years. Male:female ratio was 10:3. All the patients had fever followed by peeling (81%), redness (80%), mucosal changes (62%), conjunctival congestion (62%), rash (61%) and lymphadenopathy (35%). Family history was noted in one patient. All patients received IVIg (2gm/kg) and aspirin as standard treatment.

Estimation of CECs ($CD45^{dim}/CD146^{+}/CD31^{+}/CD133^{-}$) and EPCs ($CD34^{+}/CD309^{+}/CD133^{+}$) was done using specific antibody markers tagged with different fluorochromes, using a specific gating strategy, acquired on flowcytometer (Beckman Coulter Navios) and analyzed using kaluza software.

Results:

- Significantly elevated no. of EPCs were found in patients in group 1, 2 and 3 (P-<0.0001, P-0.0001, P-0.0003) respectively as compared to healthy controls. Higher number of EPCs were noted in patients with aneurysms [(Group 2: P-0.01, Group 1,3 (non-significant)] as compared to patients without aneurysms.
- Significantly elevated no. of CECs were noted in patients in group 1, 2 and 3 (P-<0.0001) as compared to healthy control. Also higher number of CECs was noted in group 1 and group 2 in patients with CAA as compared to patients without CAA that showed statistical significance (P-0.04, P-0.01 respectively).

Conclusion:

The initial inflammatory insult to the endothelium leads to endothelial injury, while the persistent chronic & smouldering inflammation leads to endothelial dysfunction. The endothelial dysfunction in KD patients has been found to persist for several years after the acute stage and is a potential cardiovascular risk factor. Enumeration of EPCs can be used as a good screening marker in follow up patients. Increased no. of CECs reflects endothelial damage in KD patients and may serve as early diagnostic tool for assessing endothelial damage. Higher no. of cells in KD patients without CAA

highlights their progression to endothelial damage in future if not treated. So, more close follow up is needed for these patients also as they can be at high risk of developing aneurysms.

Estimation of protein levels of inflammation induced-endothelial cell dysfunction markers in patients with Kawasaki disease from North India

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lntroduction:

Kawasaki disease (KD) is an acute self-limited vasculitis with a predilection for coronary arteries. Children with KD may have abnormal lipid profiles, altered cytokine profile and adhesion molecules, altered levels of proteins involved in vascular injury and remodeling and other inflammatory markers having associations with vascular endothelial dysfunction. The initial inflammatory insult to the endothelium leads to endothelial injury, while the persistent chronic & smouldering inflammation leads to endothelial dysfunctio

Methods:

The present study is a single centre, prospective study in North India to estimate protein levels of inflammation induced-endothelial cell dysfunction markers in patients with KD. Patients were diagnosed as per (AHA) criteria 2004 at different time intervals with and without coronary artery aneurysms (CAA) (Z score as per Montreal (JASE 2011). KD patients with CAA were further classified into transient and persistent aneurysms as per AHA 2017.

Four groups were enrolled-

Group 1: Diagnosed prior >6 months-1.5 years **(N=19)**

Group 2: Diagnosed >1.5 - 3 years **(N=24)**

Group 3: Diagnosed > 3 - 4.5 years **(N=22)**

Group 4: Age and sex matched healthy controls **(N=20)**

Maximum patients (56%) were <5 years of age while 12% were > 10 years of age. Median age was 4 (range: 0.2-13) years. Male:female was 10:3. All the patients had fever followed by peeling (81%), redness (80%), mucosal changes (62%), conjunctival congestion (62%), rash (61%) and lymphadenopathy (35%). Family history was noted in one patient. All patients received IVIg (2gm/kg) and aspirin as standard treatment. Few patients received IVIg+Infliximab (5-10gm/kg).LMWH was given in patients with giant aneurysms. Concentration of protein levels were estimated by Luminex technique (Multiplexing array based on solid-phase bead-based sandwich immunoassay) using MAGPIX with Xponent software and analysed by Belysa TM 1.2.0. Protein levels of resistin and osteopontin were measured using ELISA (Enzyme linked immunosorbent assay-Sandwich ELISA). Optical density was determined using a microplate reader (Tecan) at 450nm.

Results:

Significantly elevated pentraxin3 levels in KD patients (P-0.03, P-0.01) suggest it as a definitive biomarker for the prediction of KD. Comparable values were noted in PECAM-1 levels in patients and healthy controls in all 3 groups. Elevated VEGF-A levels were observed in KD patients. Significantly elevated levels of proinflammatory cytokine -CXCL8 and angiopoietin-2 in patients as compared to healthy controls as well as in patients with aneurysms suggesting its role in pathogenesis of KD and progression of disease in patients with CAA. Higher serum resistin levels were observed in patients as compared to healthy control while no significant difference was found in patients with and without CAA. Significantly elevated osteopontin levels were observed in KD patients.

Conclusion:

Elevated vascular endothelial growth factor-A levels involve promotion of vascular permeability so implying its possible role in development of CAL. Elevated levels of pro

inflammatory cytokine suggests its role in pathogenesis of KD and progression of disease in patients with CAA. There is disruption of vascular homeostasis in KD patients and in patients with aneurysms with altered lipid metabolism. Elevated osteopontin level is associated with increased risk for vascular injury in KD patients suggesting its role as a potential biomarker for vascular inflammatory disease.

CD40 expression and *CD40* gene polymorphism in children with Kawasaki disease

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Introduction: Kawasaki disease (KD), also known as mucocutaneous lymph node syndrome, is a medium vessel vasculitis seen predominantly in young children.KD is believed to develop in a genetically susceptible host on exposure to an environmental agent. Coronary artery abnormalities (CAAs) may develop in approximately 15-20% of patients with KD if the diagnosis is missed and treatment is delayed. Single nucleotide polymorphisms in several genes have been implicated in the pathogenesis of KD. Polymorphism of *CD40* gene has been well documented for susceptibility and development of CAAs in KD especially in Japanese, Chinese and Taiwanese population.However, there are no data on SNPs in *CD40* gene in patients with KD from Indian subcontinent. We evaluated CD40 gene polymorphism, and CD40 expression in children with KD from North India.

Aim: To study CD40 expression and*CD40* gene polymorphism in children with Kawasaki Disease as well as age matched febrile and healthy controls.

Patients and Methods: Patients diagnosed to have KD was based on the diagnostic guidelines proposed by the American Heart Association (AHA).Flow cytometric estimation of CD40 expression was estimated in different groups of the current study. The estimated percentage of CD40 expression on B cells, stimulation index (SI) and delta mean florescence index (Δ MFI) between study and control group was documented.The DNA of all the samples was amplified of *CD40* gene covering both polymorphism (*rs153045* and *rs4810485*) and ran through gel electrophoresis. Metanalysis was carried out to assess the role of both SNPs in *CD40* gene in the pathogenesis of CAAs. GRADEpro GDT software (v.3.2) was used to assess 'certainty of evidence'.

R**esults:**Forty-onepatients with KD and 41 age and sex matched healthy controls were enrolled. T allele of r*s153045 was* found to be significantly associated with KD (OR=1.28; 95% CI:1.09-1.50; p=0.002). The GRADE of evidence for this outcome was of 'low certainty'**.** However, none of the alleles and genotypes of CD40 gene were found to be associated with susceptibility to KD.The comparison of percentage expression between the groups was not statistically significant. Likewise, SI and Δ MFI of both groups were also compared. The median difference of SI and Δ MFI between KD and control group was not statistically significant (p=0.65). However, patients of KD with CAAs the median Δ MFI was noted to be higher than the febrile controls, though not statistically significant (p=0.21).

Conclusion: In the present study,SNPs *rs153045* and rs*4810485* of *CD40* gene showed no association with KD in our cohort. Secondly, the CD40 expression on B cells in acute stage of KD was also not significantly elevated, though this was higher in those with CAAs than febrile control. This could be because of a small sample size. However, metanalysis showed that T allele of *rs153045* is significantly associated with KD.

Title: Spectrum of Kawasaki Disease Spectrum in COVID Era from a single Tertiary Care Centre in Himachal Pradesh: A case series

Author Names: Dr. Ravleen Kaur, Dr. Seema Sharma, Dr. Milap Sharma

Affiliations: Department of pediatrics, Dr. Rajendra Prasad Medical college, Kangra at Tanda, Himachal Pradesh

Introduction:

Kawasaki disease (KD), a spectrum with yet unknown etiology is being increasingly reported all over India. In last few years, the vast systemic involvement in KD is being attributed to 'cytokine storm'. The current phenotype has drastically transitioned in comparison to the pre-pandemic times involving older children and atypical presentations.

Case Series:

13 children with KD between age group 1 month to 18 years were studied in period of 1 year. Clinical, lab, treatment profile and outcome were reviewed. Lab investigations including Complete blood count, ESR, CRP, Coagulation profile, ferritin, liver enzymes, LDH, Triglycerides, D dimer including SARS CoV-2 PCR and SARS CoV-2 antibody were done.

Out of KD spectrum, 3 patients presented with complete KD, 7 with Incomplete KD, 1 patient with KD with shock, 2 with Atypical KD without fever. The age group ranged from 4 months to 10 years with median age of 3 years. 8 out of 13 were male. 1 patient was a known case of B-ALL. 5 patients had GI symptoms on presentation. 2 patients had significant hepatosplenomegaly. 5 patients were critically sick and were admitted in Pediatric Intensive Care Unit. SARS CoV-2 IgG antibody was positive in 5 patients varying quantitatively from 4.6 COI to 65 COI. Coronary artery dilatation was found in 2 patients with Incomplete KD. 11 patients had raised proBNP. Ferritin were raised in all patients. Patient were started on IVIG on day 1 to day 2 of hospital stay. All patients received IVIG (Intravenous Immunoglobulin) and improved on single dose. All patients were discharged on aspirin.

Conclusion:

Early diagnosis due to increased awareness and early treatment in our Centre had good outcome. Kawasaki disease should be considered a part of hyperinflammatory syndrome with cytokine storm being the potential etiology.

Title: Diverse Spectrum of Kawasaki Disease Spectrum in COVID Era from a single tertiary care centre in Himachal Pradesh: A case series

Author Names: Dr. Ravleen Kaur, Dr. Seema Sharma, Dr. Milap Sharma

Affiliations: Department of pediatrics, Dr. Rajendra Prasad Medical college, Kangra at Tanda, Himachal Pradesh

Main Body:

Kawasaki disease (KD), a spectrum with yet unknown etiology is being increasingly reported all over India. In last few years, the vast systemic involvement in KD is being attributed to 'cytokine storm'. The current phenotype has drastically transitioned in comparison to the pre-pandemic times involving older children and atypical presentations.

Objective:

To study the profile of KD spectrum patients from Oct,2021 to Oct,2022. Clinical, lab, treatment profile and outcome were reviewed. Patient were assessed for varying presentations and outcome.

Method:

All children admitted to the Centre with KD from October,2021 to September,2022 were included .13 children with KD between age group 1 month to 18 years were studied in period of 1 year. Clinical, treatment profile and outcome were reviewed. Lab investigations including Complete blood count, ESR, CRP, ferritin, liver enzymes, LDH, Triglycerides, D dimer including SARS CoV-2 antibody were done.

Results:

Out of KD spectrum, 3 patients presented with complete KD, 7 with Incomplete KD, 1 with KD with shock, 2 with Atypical KD without fever. The age group ranged from 4 months to 10 years. (median age - 3 years). 8 out of 13 were male. 1 patient was a known case of B-ALL. 5 patients had GI symptoms on presentation. 2 patients had significant hepatosplenomegaly. 5 patients were critically sick and were admitted in Intensive Care Unit. SARS CoV-2 IgG antibody was positive in 5 patients varying quantitatively from 4.6 COI to 65 COI. Coronary artery dilatation was found in 2 patients. 11 patients had raised proBNP. Ferritin were raised in all patients. All patients received IVIG (Intravenous Immunoglobulin) and improved on single dose. All patients were discharged on aspirin.

Conclusion:

Early diagnosis due to increased awareness and early treatment in our Centre had good outcome. The presentation of KD in recent times has varied.

Title: Kawasaki Disease vs KD Phenotype Multisystem Inflammatory Syndrome- in Children (MIS-C): Two sides of the same coin.

Auhor: Dr. Ravleen Kaur, Dr. Seema Sharma, Dr. Milap Sharma

Affiliations: Department of pediatrics, Dr. Rajendra Prasad Medical college, Kangra at Tanda, Himachal Pradesh

Introduction:

Children with MIS-C have overlapping clinical features with other hyperinflammatory syndromes, both due to exaggerated immune response. A Subtype of MISC has features similar to Kawasaki Disease (KD) with more severe systemic involvement requiring different treatment protocol as compared to KD.

Objective:

To study the profile of KD and KD Phenotype MIS-C patients from Oct,2021 to Oct,2022. Clinical, lab, treatment profile and outcome were reviewed and compared.

Method:

Patients were selected on basis of AHA guidelines for KD and WHO guidelines for KD Phenotype MIS-C. Profile of all these patients admitted in our centre were assessed.13 children with KD and 6 children with KD phenotype MIS-C were studied in period of 1 year. Clinical, laboratory, treatment profile and outcome were reviewed.

Results:

The age group for KD ranged from 4 months to 10 years and 1.5 years to 12 years for KD phenotype MIS-C. GI symptoms on presentation was more common in KD in our centre. 5 KD patients and 1 patient with KD phenotype MIS-C were critically sick and admitted in Pediatric Intensive Care Unit. SARS CoV-2 IgG antibody was positive in 5 KD patients varying quantitatively from 4.6 COI to 65 COI. Coronary artery dilatation was found in 2 KD patients only. Leukopenia, lymphopenia and thrombocytopenia were more common in KD Phenotype MIS-C. Ferritin were raised in all patients. All patients with KD received IVIG(Intravenous Immunoglobulin) and improved on single dose and were discharged on aspirin. MIS-C patients received IVIG and methylprednisolone with nomortality. 1 patient in KD phenotype MIS-C received enoxaparin.

Conclusion:

It is controversial to say whether KD phenotype MIS-C is a new entity or is a severe type of KD. The likely etiology remains the same i.e cytokine storm, thus leading to more similarities than difference between the two diseases.

FEBRILE SEIZURES AS A PRESENTATION OF KAWASAKI DISEASE

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Department of Paediatrics [1]and Rheumatology[2], Yashoda Hospital,Malakpet,Hyderabad,Telangana

A 3-year-old girl child presented to emergency with a history of an episode of generalized clonic tonic seizures after a high-grade (102F) fever spike. The child had a previous history of two febrile seizures. She persisted to have fever spikes without any other symptoms or signs in the next 48 hours. On day 3, the child developed a diffuse erythematous maculopapular rash over the trunk which later spread all over the body. On examination, there was bulbar conjunctival congestion and bilateral cervical adenopathy(>1.5cm). On day 4 of illness, the child had erythema and cracking of lips, strawberry tongue, and oropharyngeal erythema present.

Hemogram showed normocytic normochromic anemia, leucocytosis and normal platelets. She had a very high ESR and CRP. The biochemical evaluation revealed hypoalbuminemia and transaminitis. Her blood and urine cultures were sterile and other infective workups (Dengue, Parasite F&V, Scrub, and typhoid) were negative. She was diagnosed with Kawasaki disease with the above clinical and laboratory features. The child was started on iv immunoglobulin(2gm/kg) along with aspirin on Day4. The child became afebrile after 48 hours. Aspirin continued for 6 weeks and stopped. Follow-up 2D Echo showed normal coronaries. Febrile seizure as a presentation of Kawasaki is rarely described. A very high index of suspicion is required for Kawasaki disease even in children with a previous history of febrile seizures as the early diagnosis and initiation of iv immunoglobulin will save a child from long-term complications of the disease.

MISC – KD (multisystem inflammatory syndrome in children-Kawasaki disease) Phenotype in SARS COVID 2ND wave – a preliminary audit

Soumita Sarkar,SanjuktaSaha,Debapriya Roy, Aniket Roy, Mihir Sarkar, Mithun Chandra Konar, Mausumi Nandi, RakeshMondal

From Pediatric Intensive Care Unit(PICU), Department of Pediatrics, Medical College Kolkata

Background and objectives: Cytokine release syndrome(CRS), which occurs in both KD as well as in MISC encompasses disorders of immune dysregulationcharacterised by constitutional symptoms, systemic inflammation andmultiorgan dysfunction. The objective ofour study is to delineate the clinico-etiological parameters, laboratory findings, treatment and outcome of children (1 month-12 years) developing MISC-KD phenotypeadmitted in PICU over a period of 6 months.

Materials and methods:A prospective observational study was doneand relevantinformations were collected. MISC criteria as defined by ICD-10 was taken as our working definition.

Results: Out of87 admission, 19.5% (n=17) fulfilling our criteria were recruited. The study population has 52% boys with median age of 6 years and mean hospital stay for 15 days. After admission 41.1% children were diagnosed with acute encephalitis syndrome, 29.4% were found to have ARDS and remaining 29.4% were found to have pyrexia of unknown origin.MISC-KD phenotype developed within 5 days of PICU stay in 41.1% (n=7).Pulmonary and neurological features were seen in 29.4 % children each.Increase in serum ferritin was seen in 45% (n=7). Among all children, 17.6% required HFOV support, 58.8% were on invasive ventilation and rest received high flow oxygen. Steroids plus antibiotics were required for managing 47% and 41% children required intravenous immunoglobulin. The mortality rate was 17% (n=3).

Conclusion: Children of all age groups can be affected but it's the younger age group that are mostly and more severely affected by COVID 19 infection.Overall the disease carries less mortality in pediatric population and like adults comorbidities and concomitant infections could alter disease outcome. Of late MISC a new challenge for pediatrician which is more common in elderly children, however it had a good prognosis with immunosuppressive therapy.

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Abstract:

Title: Increasing incidence of Kawasaki disease at Chandigarh is a fact: our experience over 26 years

Authors: Suprit Basu, Rakesh Kumar Pilania, Rajni Kumrah, Reva Tyagi, Pallavi Nadig L, Sanjib Mondal, Ankur Kumar Jindal, Pandiarajan Vignesh, Deepti Suri, Manphool Singhal, Surjit Singh

Affiliations: Postgraduate Institute of Medical Education & Research, Chandigarh, India

Title: Increasing incidence of Kawasaki disease at Chandigarh is a fact: our experience over 26 years

Background: There is a paucity of literature on epidemiological data on Kawasaki disease (KD) from the developing world. Aim of the present study is to find the incidence of KD,seasonal variations and coronary artery abnormalities(CAAs) in children with KD fromChandigarh, union territory(UT), India.

Methodology: All children with KD from Chandigarh,who presented to Pediatric Allergy Immunology unit, Postgraduate Institute of Medical Education and Research, Chandigarh, a tertiary care centre in north India, from January 1994 to December 2021were analyzed.

Results: During 1994-2019, of 1014 cases of KD, 217 (157 boys, 60 girls) were identified from Chandigarh. Mean age at diagnosis was 58 months (median=48 months; range: 12 days - 15 years). Incidence rates gradually increased during the 26 years period from 0.51 per 1,00,000 in 1994 to 10.6 per 100,000 children below 5 in 2019. Maximum number of cases were observed in the year 2019 followed by 2018. Comparative analysis revealed a progressively increasing incidence of KD from 3.1 in 1994-2008; 11in 2009–2014 to 17 in 2015-2019 per 100,000 children below 15 years. There were clustering of cases during the

months of April-May and September-November with highest peak in October followed by May, while nadir was seen in February. CAAs during the acute phase of illness were seen in 25(11.5%) cases. Frequency of CAAs during different time period were- 4 in 1994-2008; 7 in 2009-2014; 14 in 2015-2019.

Conclusions: This study highlights that the incidence of KD in Chandigarh has increased over the last 26 years. This may reflect a true increase in KD incidence or may be due to increased ascertainment of disease as a result of increased awareness among pediatricians and physicians. The seasonal pattern of KD showeda peak in October and nadir in February. Despite treatment, CAAshave been reported in as high as 11.5% of patients with KD during the acute phase of the disease.

Intravenous immunoglobulin vs Methylprednisolone in Multisystem Inflammatory Syndrome in Children with Covid 19- an observational study

Syed Ashfaq Hussainie, Mushtaq Ahmad Bhat

Abstract: The Covid 19 disease has led to a global healthcare crisis causing significant morbidity and mortality. A new disease entity known as multisystem inflammatory syndrome in children (MIS-C) has been found to be temporally associated with SARS CoV infection with a possible immune dysregulation subsequent to infection. MIS-C can present with mild symptoms like fever and dermatological involvement to severe multiorgan involvement , shock, LV dysfunction and coronary dilatation. Treatment guidelines for MIS-C have evolved from time to time since the disease became the focus of attention. We describe the clinical profile of 35 MIS-C patients admitted in our unit and compare the outcomes of pulse methylprednisolone and a combination of intravenous immunoglobulin and pulse methylprednisolone as standard therapy in moderate to severe cases.

Methods: Prospective observational study conducted in tertiary care centre in North India. Demography, clinical, laboratory variables, treatment and outcome were entered into proforma and analysed

Result: 35 cases met the inclusion criteria. 66% were males 34% females. Fever was most predominant feature (100% cases).Covid Ig was present in 100% cases , no case had Covid RTPCR positive. 67% had moderate MIS-C and 33% had severe MIS-C. 42.9% cases had LV dysfunction. 19% cases had coronary dilatation. 2 patients had prolonged PR interval and 1 patient had sinus nodal dysfunction. Higher age(correlcoeff 0.65), HsCRP(correl=0.545), ESR(correl=0.109), serum ferritin(correl=0.50),D-dimer(correl=0.42), IL6(correl=0.83) were associated with severe illness. 48% cases received only pulse methylprednisolone , 52% cases received combination of IVIG and pulse methylprednisolone. Combination therapy had better defervescence compared to methylprednisolone only (p value <0.05) and better improvement in LV dysfunction (p value<0.05)

MIS-C presenting as a surgical emergency in children

Syed Ashfaq Hussainie, Mushtaq Ahmad Bhat

Abstract:

The Covid 19 disease has led to a global healthcare crisis causing significant morbidity and mortality. A new disease entity known as multisystem inflammatory syndrome in children (MIS-C) has been found to be temporally associated with SARS CoV infection with a possible immune dysregulation subsequent to infection. MIS-C can present with mild symptoms like fever and dermatological involvement to severe multiorgan involvement , shock, LV dysfunction and coronary dilatation. Unusual presentations of MISC have also like stroke, encephalitis , muscle spasms have been reported. We describe 4 cases with unusual presentation of MIS-C. Three patients presented with features mimicking acute abdomen and one patient presented with acute kidney injury.

Method: Case series conducted in a tertiary care centre in North India. Demography, clinical, laboratory variables, treatment and outcome were entered into proforma and analysed.

Results: Four patients 1 girl and 3 boys. Mean age 8.75 years(SD=2.75). Initial complaint was fever in all cases. Two patients had pain abdomen mimicking acute appendicitis. One underwent negative laparotomy for suspected appendicitis, other was managed conservatively. Third patient had abdominal distension and constipation mimicking subacute intestinal obstruction. Fourth patient had acute kidney injury (creatnine 4.5, urea 180) needing dialysis. Mean WBC count was 9925(SD=9184.18), mean platelet count was 389250(SD=312733.5), mean HsCRP was 135(SD=45.6), mean ESR was58(SD=11.22), mean ferritin level was 694(SD=127.3), mean D dimer was 5285(SD=4876), mean pro BNP level was 13450(10246.78). All patients responded to a combination of Intravenous Immunoglobulin and pulse methylprednisolone.

Long term Consequences of delayed/denied treatment of Kawasaki disease in infants

Mushtaq Ahmad Bhat

Kawasaki disease is a medium vessel vasculitis of young infants and children. Delayed or denied treatment can lead to coronary artery aneurysms which can have devastating long term consequences for these children.

We present long term follow up of two cases of Kawasaki disease diagnosed in infancy who inspite of presenting early were either diagnosed late or treatment was delayed

Patient A, a 7 month old infant presented to our hospital with history of fever and rash for 5 days in 2013. Initial diagnosis of KD was made on day 2 of admission .However it took another 5 days to decide about the therapy with IVIG. He received IVIG on day 10 of fever. Fever reappeared after 48 hours. Second dose of IVIG could not be given due to confusion regarding the diagnosis and denial of the disease and ultimately patient was discharged after next 3 days. During follow-up patient developed desquamation of skin and Bues lines during 3^{rd} to 4^{th} week. Subsequently developed multiple coronary artery aneurysms and over 7 years has developed dilatation of aortic root.

Patient B was admitted at 6 months of age with history of fever for 7 days. No treatment was give for next 5 days as all investigations for infections were normal Before discharge on day 12 an ECHO was done which showed aneurismal dilatation of coronary arteries. Patient received IVIG and infliximab. However he developed aneuryms of all three branches of coronary arteries. After 5 years of follow-up he continues to have aneurysms of 2 branches of coronary arteries.

In conclusion delayed or denied treatment of patients with KD can lead to aneurismal dilatation of coronary arteries which may persist and can have disastrous consequences for these poor kids.

Prevalence and predictors of Macrophage Activation Syndrome in Kawasaki Disease: A single center observational study

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Background: Beginning of the SARS-CoV-2 pandemic was marked by an increase in the incidence of Kawasaki Disease (KD) and its antecedent complications like Macrophage activation syndrome (MAS), overt myocarditis and Kawasaki Disease Shock Syndrome (KDSS) [1].

Objectives: We aimed to study the prevalence and the early predictors of MAS in patients with KD

Methods: All childrenin the age group of 1-18 years admitted in our tertiary care hospital during the period of March 2020 to October 2021 who were diagnosed with KD as per AHA 2017 and had a negative SARV-CoV-2 serology/ RTPCR were enrolled [2]. All patients were investigated for MAS as per International Consensus criteria by Ravelli*et al* [3]. A comparative analysis of clinical and laboratory parameters was done in KD patients with and without MAS. Logistic regression analysis was done for predicting the odds of MAS in KD and a classification tree was built to predict MAS in KD.

Results: Atotal of42 patients were enrolled in the study out of which 14 (33.3 %) were diagnosed with MAS. The KD patients with and without MAS did not show any significant difference in the clinical phenotype as shown in Table 1. As per multivariate logistic regression analysis, children with underlying myocardial dysfunction, hypoalbuminemia of <2.8 gm/dl, duration of fever >6 days were strong predictors of MAS.The classification tree generated by modelling is shown in Figure 1

Conclusion: MAS remains an under-reported entity with grave outcomes in patients with KD due to a significant clinical overlap. There has been a surge in the prevalence of inflammatory disorders including KD and associated MAS reported during the SARS-COV2 pandemic. Earlyscreening for MAS using easily available bedside parameters can help in early detection and timely institution of specific therapy and improve disease outcomes.

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Table 1: Clinical profile and laboratory parameters of KD patients with and without MAS

Clinical Characteristics	**MAS (n= 14)**	**Non-MAS (n= 28)**	**P value**
	Median (IQR) / frequency (%)	**Median (IQR) / frequency (%)**	
Age (months)	48 (2-72)	24 (12-72)	0.7
Gender Male Female	 11 (64.7%) 06 (35.3%)	 17 (68%) 08 (32%)	1
Duration of fever (days)	07 (5-15)	05 (5-9)	**0.05**
Rash	10 (58.8%)	15 (60%)	1
Mucositis	08 (47%)	12 (48%)	1
Lymphadenopathy	03 (17.6%)	06 (24 %)	0.72
Eye redness	11 (64.7%)	11 (44%)	0.32
Desquamation of the skin	05 (29.4%)	03 (12%)	0.23
Extremity edema	08 (47.1%)	11 (44%)	1
Complete Kawasaki disease	06 (35.3%)	07 (28%)	0.87
Incomplete Kawasaki disease	11 (64.7%)	18 (72%)	0.87
Kawasaki disease with infection	08 (47%)	13 (52%)	1
Organ dysfunction			
Requirement of oxygen	09 (52.9%)	07 (28%)	0.19
Shock requiring inotropes	11 (64.7%)	11 (44%)	0.32
Acute kidney injury	06 (35.3%)	03 (12%)	0.12
Liver dysfunction	09 (52.9%)	07 (28%)	0.19
Myocarditis	10 (58.8%)	07 (28%)	**0.09**

Coronary Artery Aneurysm	03 (21.4%)	04 (14.2%)	0.4
CNS	03 (17.6%)	01 (4%)	0.29
Laboratory parameters			
Hemoglobin (g/dl)	9.7 (7.9-10.45)	9.5 (7.9-10.25)	0.48
Total leukocyte count (x10 9/L)	11200 (7690-25000)	13370 (8600-18300)	0.59
Platelet count (x10 9/L)	1.4 (1.00-5.9)	2.3 (1.27-2.60)	0.75
CRP (mg/L)	114 (56-205)	130 (60-213)	0.64
D-dimer (ng/ml)	1980.5 (768.5-2150)	1068.5 (555.75-1915.75)	0.75
Fibrinogen (mg/dl)	378 (248-446)	390 (346-466)	0.32
Ferritin (ng/mL)	1200 (833-1500)	189 (167-230)	7.8E-08
Albumin (g/dl)	2.3 (2.2-2.7)	2.8 (2.3-3.1)	0.03
Triglyceride (mg/dl)	136 (123-212)	134 (116-177)	0.69

Abstract:

Title: Study of association of $F_c\gamma$Rs in patients with Kawasaki Disease

Introduction: Kawasaki disease (KD) is an acute febrile systemic vasculitis that affects coronary arteries. Administration of high dose intravenous IVIg provides a prompt anti-inflammatory effect through mechanisms that are not clearly understood. Clearance of autoantibodies on binding with F_c receptors, provision of the anti-idiotypic antibodies, blockade of adhesion molecules and activation of the inhibitory F_c receptor (i.e. $F_c\gamma$RIIB) on macrophages are some proposed mechanisms. Altered FcγR expression may result in unbalanced immunity or auto-inflammation. FcγRreceptors are suggested to have important role in mechanism of IVIg action as well as in IVIg resistance, development of coronary artery aneurysms (CAAs) and myocardial dysfunction that occurs in patients with KD. In recent Genome Wide Association Studies (GWAS) studies, a functional variant in FCGR2A (131H>R; rs1801274) was associated with susceptibility of KD.FCGR2A, located within the FCGR2/3 gene cluster encodes the Fc-gamma receptor (FcγRIIa, an activatory receptor).This cluster contains extensive gene copy number variations (CNVs) and several important single nucleotide polymorphisms (SNPs). SNPs and CNVs in the FCGR locus may be a regulating factor for gene transcription and expression of the FcγRs, and hence for susceptibility to KD.

The current study is designed to characterize the surface expression of $F_c\gamma$Rs ($F_c\gamma$RI, $F_c\gamma$RII, $F_c\gamma$RIII) in patients with KD and their polymorphisms in Indian population. A better understanding of these pathways are likely to open up better therapeutic options in patients with KD especially IVIg resistance thereby prevent or control the progression of cardiac diseases.

Material and Methods:

Treatment naïve children with KD were enrolled in group I. Group II included children post treatment withIVIg. They were classified as IVIg responders or IVIg resistant KD based on persistence of fever at 36 hrs of IVIg. In addition, 15 healthy controls were also enrolled. Clinical and laboratory profiles of patients were obtained from hospital records. Characterization of surface expression of $F_c\gamma$Rs was done using flow cytometry. Genetic polymorphisms were determined with an FCGR-specific Multiplex Ligation-dependent Probe Amplification (MLPA) assay (MRC-Holland, Amsterdam, The Netherlands).

Results: 15 children were enrolled in group I and II. CD16 ($F_c\gamma$RIII) and CD32($F_c\gamma$RII) levels were reduced in patients as compared to healthy controls. Levels of $F_c\gamma$Rs were downregulated in post IVIg patients as compared to pre IVIg patients.A few SNPs have been associated rs1050501 (*FCGR3B*, p.Ile232Thr), rs1801274 (*FCGR2A*; p.His166Arg); rs396991 (FCGR3A, p.Val176Phe) in Indian population. The latter two are associated to increased affinity for human IgG binding. Variants in 3 children were identified in these patients.CNVs in CNR4 have also been identified in our population.