

# THE LANCET

## Rheumatology

### Supplementary appendix

This appendix formed part of the original submission. We post it as supplied by the authors.

Supplement to: Vergnano S, Alders N, Armstrong C, et al. Severe refractory Kawasaki disease in seven infants in the COVID-19 era. *Lancet Rheumatol* 2020; published online July 10. [https://doi.org/10.1016/S2665-9913\(20\)30231-9](https://doi.org/10.1016/S2665-9913(20)30231-9).

## Appendix 1

Table 1 Demographic and clinical characteristics of the infants

	Infant 1	Infant 2 **	Infant 3	Infant 4	Infant 5	Infant 6	Infant 7
Age	5 months	7 months	7 months	12 months	12 weeks	5 months	4 months
Sex	Male	Male	Male	Male	Female	Female	Male
Ethnicity	White not British	White British	Asian	Mixed race	White British	White British	Asian
Symptoms at diagnosis							
Fever	For 4 days	Fever 3 days	For 8 days	For 5 days	For 5 days	For 5 days	For 5 days
Rash	Generalised macular erythematous	Generalised erythematous	Maculo-papular erythematous generalised	Maculo-papular erythematous generalised	Discrete macular blanching on torso	Intermittent macular rash, generalised	Polymorphous erythematous rash
Peeling of hands/feet	Yes	Yes	Yes	No	No	No	No
Bilateral not exudative conjunctivitis	Yes	Yes	Yes	Yes	Yes	No	Yes
Lymphadenopathy	No	No	No	No	No	No	Yes
Strawberry tongue/red lips/pharyngitis	Yes	Yes	Yes	No	No	Yes	Yes
Oedema/periungual peeling of hands or feet	No	Yes	Yes	Yes	No	No	No
Prominent BCG site	Not vaccinated	n/a	Recent swelling and scabbed	Yes	Not vaccinated	Not vaccinated	Not vaccinated
Other	Very irritable	None	Vomiting, diarrhoea and cough	Miserable/irritable	Diarrhoea lethargic	Diarrhoea	None
Complete Kawasaki *		Yes	Yes				Yes
Incomplete Kawasaki	Yes			Yes	Yes	Yes	
Investigations							

CXR	Normal				R upper lobe changes	Normal	Mild peribronchial thickening
BC	Negative	Negative	Negative	Negative	Negative – CoNS contaminant	Negative	Negative
CSF	Negative (incl virology)	Not done	Not done	Not done	Negative (incl virology)	Not done	Negative (incl virology)
Urine	Negative		Negative		Not done		Negative
ASOT and antiDNase	Negative	Negative	Negative	Negative	Not done	Not done	Not done
COVID 19 PCR	Not done	Negative	Negative	Positive	Negative	Negative	Negative
COVID 19 serology	Negative	Negative	Negative	n/a	Negative	Positive	Negative
Respiratory viral panel	Negative	Adenovirus positive	Negative	Negative	Negative	Negative	Rhinovirus positive
EBV serology	Negative	Negative	Not done	Negative	Not done	Not done	Not done
CMV	Negative	Negative	Negative on respiratory PCR	Negative	Not done	Not done	Not done
Stool virology	Negative	Not done	Negative	Not done	Not done	Not done	Not done
Echocardiogram							
Coronary arteries	Bilateral coronary artery dilatation	Bilateral giant coronary artery aneurysms	Left coronary artery aneurism	RCA aneurism	Normal	Bilateral giant coronary artery aneurysms	LAD mild dilatation
Z score at discharge	RCA: + 7.42 LMCA + 9.41 LDA	RCA coronary + 20 LMCA +29 LDA + 50	RCA +2.2 LMCA +5.2 LAD+17.5	RCA +5.50 LMCA +4.18 LAD +2.43	RCA +1.6 LMCA +1.05 LAD +1.52	RCA +20 LCA +23	RCA +2.2 LMCA +0.89 LAD +4.14
Cardiac function	Normal	Normal	Normal	Normal	Normal	Normal	Normal
Treatment							
IVIG doses	2 (day 4 and day 5 of fever) 2 g/kg	2 (day 3 and day 7 of fever)	1 (day 8 of fever)	2 (day 5 and day 7 of fever)	2 (day 5 and 5) 2 g/kg	1 (day 5) 2g/kg	1 (day 5)
Methylprednisolone	30mg/kg for 3 days (day 5 of fever)	20mg/kg for 3 days given twice (day 11 of fever and day 18 from onset of illness)	0.8mg/kg BD for 7 days (day 8 of fever)	2mg/kg/day from day 7	10mg/kg day 5 for 5 days	2mg/kg (day 7 then 10mg/kg on day 10)	10mg/kg (day 6 for 3 days)
Oral steroid course	Yes				Yes	Yes	Yes

Biological agent	Infliximab - 5mg/kg (day 10 of fever and day 45 from onset of illness)	Infliximab - 5mg/kg (day 20 from onset of illness)	Infliximab - 6mg/kg (Day 15 of fever)	Infliximab - 5mg/kg (day 8 of fever)	Anakinra- 2mg/kg BD on day 6 for 3 days	Anakinra - 2mg/kg BD (day 7) Infliximab - 5mg/kg Day 12	Infliximab - 3mg/kg (Day 8 of fever)
Aspirin	High dose initially then 5 mg/kg	High dose initially then 5 mg/kg	High dose initially then 5 mg/kg	High dose initially then 5 mg/kg	High dose initially	5mg/kg on day 5	High dose initially then 5 mg/kg
Low molecular weight heparin	Yes	Yes	Yes	Yes	No	Yes	No
Warfarin	No	No	Yes	No	No	Yes	No
Clopidogrel	No	Yes	Yes	No	No	No	No

\* Complete Kawasaki Fever for at least 5 days, and <sup>3</sup> 4 of 1. strawberry tongue or red lips or redness of oral and pharyngeal mucosa; 2.Rash; 3.Bilateral non exudative conjunctivitis; 4. Cervical lymphadenopathy >1.5 cm of diameter; Erythema/oedema or periungual peeling of hands and feet \*\* Infant died

**Figure 1 a and b** – a. Unfixed heart showing a giant aneurysm of the left main stem and left anterior descending coronary arteries. Area of perforation labelled with arrow.  
b. Left anterior descending coronary artery showing vasculitis without significant fibrinoid necrosis. The remains of the vessel wall are visible on the right (with a pink rim) with a large saccular aneurysm present on the left of the image [H&E, x25].



