Table S1. Demographic and laboratory features of the study population.

	Study group (range)	Control group (range)	p-value
Gestational age (weeks)	38,5 (28 – 42)	38,6 (25 – 42)	n. s.
Male/female (%)	37%/63%	42%/58%	n. s.
Syndromatic form (%)	10%	12%	n. s.
Age at KPE (days)	60 (26 – 142)	65 (16 – 150)	n. s.
AST at KPE (IU/l)	238 (50 – 1307)	234 (40 – 1059)	n. s.
ALT at KPE (IU/l)	162 (25 – 654)	157 (20 – 662)	n. s.
GGT at KPE (IU/l)	549 (66 – 2164)	549 (66 – 1470)	n. s.
Bilirubin at KPE (µmol/l)	136 (70 – 244)	156 (66 – 347)	p = 0.009

Table S2. Outcome related to the age in days, when the Kasai procedure was performed.

Age at KPE (days)	SOA	SNL	SNLjf
< 31	83% (10/12)	25% (3/12)	17% (2/12)
31-60	91% (68/75)	52% (39/75)	48% (36/75)
> 60	87% (64/74)	37% (27/74)	30% (22/74)

Table S3. Compilation of 14 adjuvant therapy studies after Kasai-portoenterostomy.

Author	Study design and OEBM	Patients/controls	Follow up rate % months	Drug Administration	Outcome parameters	Survival with native liver	Incidence LTx	Conclusion and comments
Meyers (2003) [1]	R IIIb	14/14	96.3% 42 months	(Methyl)prednisolone i.v. and p.o. (1)	SNL (3 -4 m) IncTx (12 m)	improved	reduced	steroids may improve the clearance of jaundice
Kobayashi (2005) [2]	R IIIb	51/12	n.a.	Prednisolone i.v. (2)	n.i.	n. i.	n. i.	not analysable
Escobar (2006) [3]	R IIIb	21/22	88.4% n.i.	Prednisolone i.v. (3)	CoJ (6 m) incLTx	improved	not reduced	significantly improved clearance of jaundice after 6 months
Vejchapipat (2007) [4]	R IIIb	33/20	n.i.	Prednisolone p.o. (4) oral	SNL (6 m)	not improved		no effect
Davenport (2007) [5]	P Ib	36/37	94.4% 12 months	Prednisolone p.o. (5) oral	jfSNL (6m and 12)	not improved	not reduced	initially improved clearance of jaundice
Petersen (2008) [18]	P IIb	49/20	100% 24 – 66 months	(Methyl)prednisolone i.v. p.o. (6)	SNL (6m and 24 m)	not improved	not reduced	high dose steroids do not reduce LTx incidence
Chung (2008) [6]	R IIIb	13/17	n.i.	Prednisolone p.o. (7)	SNL and incLTx (3 and 6 m)	not improved	not reduced	initially improved clearance of jaundice
Davenport (2013) [7]	P Ib	62/91	100% 24 months	Prednisone p.o. (8)	jfSNL (6m) SNL (48 m)	not improved	not reduced	iInitially improved clearance of jaundice

Bezerra (2014) [8]	P, DR, PC Ib	70/70	90.7% 24 months	(Methyl)prednisolone i.v. and p.o. (9)	jfSNL (6 m) SNL (24 m)	not improved	not reduced	High dose steroids do not reduce LTx incidence
Tanaka (2019) [9]	R IV	16/?	n.i.	(Methyl)prednisolone i.v. and p.o. (10)	CoJ 3m	n.i.	n.i.	not analysable
Kumar (2019) [10]	R IV	79/?	87.1% 24 months	Prednisolone p.o. (11)	CoJ (6m)	n.i.	n.i.	not analysable
Parolini (2019) [11]	R IIIb	8/28	100% 6 months	Ganciclovir Valganciclovir i.v. and p.o. (12)	SNL (24 m)	n.i.	n.i.	no clear message
Mack (2019) [12]	P IIb	29/64	96.7% 12 months	Immunoglobulines i.v. (13)	incLTx (12 m)	not improved	not reduced	no improvement of SNL with immunoglobulines
Pietrobattista (2020) [9]	P IIb	25/18	100% 24 months	(Methyl)prednisolone i.v. and p.o. (14)	SNL (24 m)	not improved	not reduced	no effect
This study	R IIb	107/83	100% 6, 12, 24, 122 and 229 months	Budesonide rectally	SNL, jfSNL (6m, 24m, 122 m, 229 m)	improved	reduced	Significantly reduced need for LTx

Study design: R - retrospective; P – prospective; RD – randomized; PC – placebo controlled. OEBM: Oxford Centre for Evidence Based Medicine, available at: http://www.cebm.net. Accessed December 1, 2020. Administration: i.v. – intravenously; p.o. orally; n.i. no information. Outcome parameters: SNL – survival with native liver (months after Kasai); jfSNL – Jaundice free survival with native liver (months after Kasai); incLTx - incidence LTx; CoJ - Clearance of jaundice (not defined). Dosage and application:

(1) Tapering 10, 8, 6, 5, 4, 3, 2 mg/kg/d (1 week), follow by prednisone p.o. 2 mg/kg/d for 8 – 12 weeks; (2) 4 different protocols (P), each dose was given for 3 days: P1: 6, 4, 2mg, P2: 10, 5, 2mg, P3: 20, 15, 10, 5, 2mg, P4: same as P3 + each time stools turn pale, protocol was restarted at 20mg; (3) Doses varied from 2 mg/kg/d (low) to 20 mg/kg/d (pulses) tapering over 2 – 6 weeks; (4) 4 mg/kg/d for 3 to 4 days, then at alternate days for 1 – 3 months; (5) 2mg/kg/d day 7 to 21and 1mg/kg/d day 22 to 28; (6) Methylprednisolone i.v. 10mg/kg/d day 1 to 5 followed by Methylprednisolone p.o. 1mg/kg/d day 26 to 28; (7) Prednisolone p.o. 4mg/kg/d week 2 – 3, 2mg/kg/d week 4 – 5, 1mg/kg/d week 6 – 7; (8) Low dose regimen: Prednisolone p.o. 2mg/kg/d day 7 to 2, 1mg/kg/d day 22 to 28, High dose regimen: Prednisolone p.o., 5mg/kg/d day 5 to 9, 4mg/kg/d day 10 to 14, 3mg/kg/d day 15 to 19, 2mg/kg/d day 20 to 24, 1mg/kg/d day 25 to 29, then Hydrocortisone: 2.5mg/kg twice per dayfrom day 30 to 32 and once per day from day 33 to 35; (9) Methylprednisolone i.v.4mg/kg/d for 2 weeks,followed by prednisolone p.o. 2mg/kg/d for 2 weeks, followed by a 9 week tapering protocol; (10) Prednisolone p.o. 4mg/kg/d for 5 days, 2mg/kg/d for 5 days, 1mg/kg/d for 5 days, 0.5mg/kg/d for 5 days, pulse therapy: methylprednisolone i.v. 20mg/kg/d for 2 days, 10mg/kg/d for 2 days, 5mg/kg/d for 2 days, followed by an oral prednisolone protocol; (11) Prednisolone p.o. 2mg/kg/d for 2 weeks, followed by a 4 week tapering protocol; (12) Ganciclovir i.v. 5mg/kg/d for 1 week, followed by valganciclovir p.o. 520mg/m2/d until negativity of CMV DNA titer; (13) immunoglobulines i.v. 1g/kg IVIg at days 3 to 5, 30 and 60 days after KPE; (14) Methylprednisolone i.v. 10mg/kg/d, decreasing 2mg/kg/d every 24h, until the dose of 2mg/kg/d, followed by prednisolone p.o. 2mg/kg/d for 15 days, 1mg/kg/d for 15 days, 0.5mg/kg/d for 15 days, 0.25mg/kg/d for 15 days

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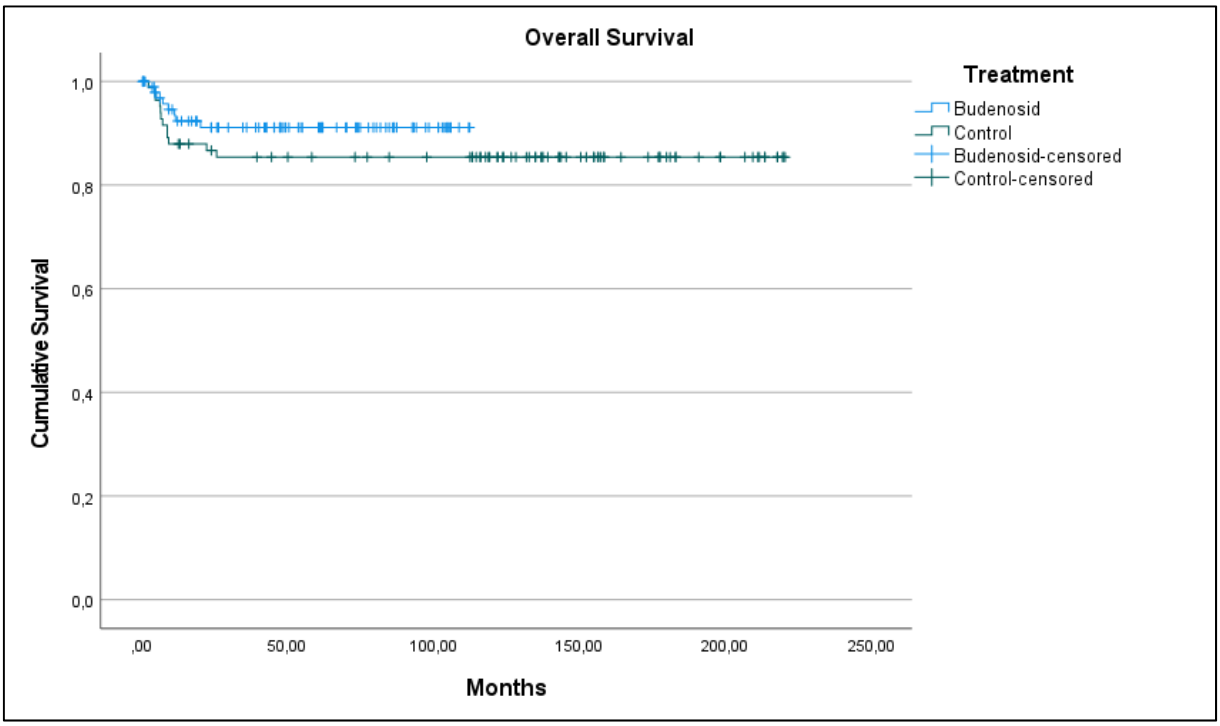

Figure S1. Kaplan-Meier curves: **s**urvival over all.